Echocardiographic Assessment of RV Function and Failure in Children with Pulmonary Hypertensive Vascular Disease

Mark K. Friedberg, MD

Outline

• Conventional Doppler
• 2-D assessment of RV function
• Annular motion
• Assessment of myocardial deformation
• Isovolumic acceleration
• The duration of systole and diastole
• Capacitance

Clinical questions to be answered by echo:

• Severity of pulmonary hypertension (Pulmonary artery systolic and diastolic pressures)
• Underlying lesions / shunting across intra-cardiac communication
• Associated lesions
• Right ventricular hypertrophy
• Tricuspid regurgitation
• Right and left ventricular function

Assessment of RV function is difficult

• No axis of symmetry: complicates geometric modeling.
• Endocardial delineation is difficult (prominent endocardial trabeculations)
• Retrosternal location limits acoustic access.
RV structure

Which is worse?

‘Eyeball’ assessment still most common method

RV does not tolerate acute increases in afterload

MacNee W. Am J Respir Crit Care Med. 1994;150: 833–852.
Adaptation to increased afterload

15 year old girl with presenting with pulmonary emboli

RV dilatation as initial response to increased afterload

Courtesy Luc Mertens

Lopez, J Am Soc Echocardiogr 2010;23:465-95

Parameters of longitudinal function

Tricuspid annular plane systolic excursion (TAPSE)

Koestenberger, JASE 2009
TAPSE correlates with RV EF

Deviation of TAPSE values from mean age-related reference values

López-Candales, Am J Cardiol 2006;98:973

Koestenberger, Congenit Heart Dis. 2012;7:250

TAPSE correlates with stroke volume

Ability of TAPSE to predict RV indexed stroke volume of 29 ml/m².

Forfia, Am J Respir Crit Care Med 2006 174:1034–1041

TAPSE correlates with survival

Deviation of TAPSE values from mean age-related reference values

Forfia, Am J Respir Crit Care Med, 2006 174:1034

Tricuspid tissue Doppler velocities

Eidem, J Am Soc Echocardiogr 2004;17:212-21
TDI severe PAH

Tissue Displacement

Pre-tricuspid Eisenmenger

13y, iPAH, PAH worsening, pneumonia

Normal TAPSE  Low FAC% due to apical dysfunction
Assessment of regional RV function by strain

Dambrauskaite, J Am Soc Echocardiogr 2007;20:1172

Fractional area of change


3-D assessment of RV volumes and EF

LV function influences TAPSE

López-Candales, AJC 2006;98:973
Other important 2-D parameters

Right atrial size
Pericardial effusion

Conventional echo parameters in children with PAH

Assessment of myocardial function

Kassem E, Am Heart J. 2013;165:1024-31
Strain and strain rate

Strain correlates with stroke volume

*SV determined at cath

Urheim, AJC 2005;96:1173

RV myocardial function

Isovolumic acceleration is a relatively load independent measure of RV function

Vogel, Circulation 2002;105:1693-1699
IVA is relatively load independent

Urheim, AJC 2005;96:1173

Table 2
Invasive hemodynamic and echocardiographic data before and after epoprostenol infusion

<table>
<thead>
<tr>
<th>Variable</th>
<th>Before Epoprostenol (n = 12)</th>
<th>Peak Epoprostenol (n = 12)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV peak systolic pressure (mm Hg)</td>
<td>74.5 ± 20.0</td>
<td>65.7 ± 19.3*</td>
</tr>
<tr>
<td>Pulmonary vascular resistance (Wood units)</td>
<td>18.3 ± 12.2</td>
<td>11.4 ± 10.3*</td>
</tr>
<tr>
<td>RVSVI (mL/m²)</td>
<td>27.1 ± 13.5</td>
<td>35.7 ± 15.2*</td>
</tr>
<tr>
<td>Isovolumic myocardial acceleration (m/s²)</td>
<td>1.6 ± 1.9</td>
<td>1.6 ± 1.4</td>
</tr>
<tr>
<td>RV peak systolic velocity (cm/s)</td>
<td>5.0 ± 1.9</td>
<td>6.1 ± 2.2*</td>
</tr>
<tr>
<td>RV peak systolic displacement (mm)</td>
<td>8.8 ± 4.5</td>
<td>11.1 ± 5.2*</td>
</tr>
<tr>
<td>RV peak systolic strain rate (1/s)</td>
<td>-1.2 ± 0.9</td>
<td>-1.5 ± 0.8</td>
</tr>
<tr>
<td>RV peak systolic strain (%)</td>
<td>-13.8 ± 8.5</td>
<td>-17.6 ± 9.1*</td>
</tr>
</tbody>
</table>

*p ≤0.05.

Back to Doppler....

RV strain in pediatric iPAH patients

Submitted
Doppler hemodynamics

MPI for RV function

Problems with MPI

- Cannot assess RV inflow and outflow simultaneously
- HR has to be similar in both measurements
- ICT is short in the normal RV -> IRT becomes more important (itself an independent index)
- Measurement of small intervals-prone to measurement error

The S/D Duration ratio

Eidem, Am J Cardiol 2000;86:654

Friedberg, JASE, 2006;19:1326

* a is duration of tricuspid valve regurgitation
* b is RV ejection time
SD ratio in the RV in HLHS

- **Friedberg, J Am Soc Echocardiogr 2007;20:749**

SD ratio in PAH

- **Alkon, Am J Cardiol 2010;106:430**

Ventricular interactions in PAH

- **Gan, Am J Physiol 2006; 290:1528**

The S/D duration ratio in PAH

- **Marcus, J Am Coll Cardiol 2008; 51:750**
- **Alkon, Am J Cardiol 2010;106:430**
S/D ratio=2.3

S/D ratio=1.3

13y, iPAH, pneumonia, PAH worsening

Echo predictors of mortality in Eisenmenger’s

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Odds Ratio (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>A' (ms⁻¹)</td>
<td>0.95 (0.92-0.98)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>S' (ms⁻¹)</td>
<td>1.04 (1.01-1.07)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>I' (ms⁻¹)</td>
<td>1.03 (1.00-1.07)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>E/A ratio</td>
<td>0.90 (0.80-1.03)</td>
<td>0.10</td>
</tr>
<tr>
<td>TAPSE (cm)</td>
<td>1.63 (1.36-2.02)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>EF (%)</td>
<td>0.82 (0.63-1.06)</td>
<td>0.15</td>
</tr>
</tbody>
</table>

Moceri, Circulation. 2012;126:1461-1468

Ventricular-vascular interactions

The Prognostic Value of Pulmonary Vascular Capacitance Determined by Doppler Echocardiography in Patients with Pulmonary Arterial Hypertension

Srijay Mahapatra, MD, Rick A. Nicholls, MD, Jeo K. Oh, MD, and Michael D. McGoon, MD, Rochester, Minnesota

Highest capacitance group

Middle two capacitance groups

Lowest capacitance group

Alive (%)

Death (%)

Stroke Volume

Pulmonary Artery Systolic Pressure

J Am Soc Echocardiogr 2006; 19:1045
Pulmonary arterial capacitance

\[ \text{PAC} = \frac{\text{RV stroke volume}}{\text{pulmonary artery pulse pressure}} \]

Sajan, Am Heart J 2011;162:562

Summary

- Assessment of RV function is important in PAH.
- Echo is the mainstay of imaging in PAH, but has important limitations.
- Newer methods can be combined with conventional assessment to provide more comprehensive evaluation.
- Further validation of the newer methods in clinical practice is needed.
- Left ventricular function and ventricular-vascular interactions should be accounted for.
Transesophageal echocardiography

- Can be performed relatively safely and is usually well tolerated (even when baseline oxygen saturation levels are < 80%).
- May be useful for detecting atrial septal defects / patent ductus arteriosus and for imaging pulmonary veins.
- Should be performed in lung transplant candidates (detect unsuspected intracardiac defects/shunts, proximal pulmonary artery thrombus).

SD ratio in PHTN

A higher S/D ratio is associated with:

- higher systolic, diastolic and mean PAP
- higher RVSP, RV/BP ratio
- higher PVR after exposure to 40ppm NO
- lower RV FAC%, lower RVEF (MRI)
- higher RVED
- lower systemic blood pressure
- shorter 6 minute walk distance
- younger age at diagnosis
- non-repaired CHD

What are we trying to capture?

3D assessment of RV function


RV VVI

A smaller TAPSE may originate from RV mechanical delay

Because the motion of the base of the heart plays a central role in its filling and emptying, we developed an original method to characterize the base motion dynamics throughout each cycle by use of pulsed Doppler echocardiography. A 100 Hz wall filter and low gain settings were used to record the low-frequency, high-energy Doppler signals generated by the motion of the base. From the apical four-chamber view, the sample volume was placed at the lateral margin and at the common septal margin of the tricuspid and mitral annuli.

“The method reported in the present study allows a more informative noninvasive quantitation of the cardiac base motion derived from measurements of its velocity, excursion, and acceleration.”

Table 1

Echocardiographic measurements used in pulmonary hypertension [2]

<table>
<thead>
<tr>
<th>Qualitative assessment</th>
<th>Additional echocardiographic measures described in the literature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlarged right atrium and ventricle*</td>
<td>RV end-diastolic area index</td>
</tr>
<tr>
<td>Right ventricular hypertrophy*</td>
<td>Right atrial end-diastolic area index</td>
</tr>
<tr>
<td>D-shaped left ventricular cavity with flattening of the interventricular septum in systole*</td>
<td>RV area change (percentage)</td>
</tr>
<tr>
<td>Diminished/absent atrial wave of pulmonary valve</td>
<td>Eccentricity index</td>
</tr>
<tr>
<td>Mitral systolic closure or notch in pulmonary valve</td>
<td>Severity of TR/Right atrial ratio</td>
</tr>
<tr>
<td></td>
<td>Escape diastolic</td>
</tr>
<tr>
<td>Right atrial outflow tract flow acceleration time</td>
<td>Pericardial effusion size</td>
</tr>
<tr>
<td>Right ventricular systolic flow acceleration time</td>
<td>Doppler index of global RV dysfunction as suggested by Tei et al</td>
</tr>
<tr>
<td>Right ventricular ejection time</td>
<td></td>
</tr>
<tr>
<td>Right ventricular index of myocardial performance</td>
<td></td>
</tr>
<tr>
<td>Timing of mid systolic deceleration of right ventricular ejection</td>
<td></td>
</tr>
<tr>
<td>Right ventricular long axis function (marker of overall right ventricular systolic function)</td>
<td></td>
</tr>
</tbody>
</table>

* Minimal data set required for echocardiographic evaluation of pulmonary arterial hypertension.

What should we measure?

Nelson Schiller and Annular excursion

RV strain is influenced by afterload

Weidemann, Ped Cardiol, 2002;23:292

Normal RV TDI/ strain values in children


TAPSE correlates with RV EF

López-Candales, Am J Cardiol 2006;98:973
Lissin, Am J Cardiol 2004;93:654–657

TGA after ASO with PHT

López-Candales, Am J Cardiol 2006;98:973
Lissin, Am J Cardiol 2004;93:654–657
TGA after ASO with PHT

REDUCED APICAL FUNCTION

Improved LV filling after relief of RV outflow obstruction

Lurz, EHJ 2009 30, 2266–2274

Lurz, EHJ 2009 30, 2266–2274