Imaging Assessment of the Pulmonary Valve in Stenosis/Atresia and Regurgitation

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SCAI Fall Fellows Course 2014
Las Vegas
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As a faculty member for this program, I disclose the following relationships with industry:


None
Anatomy

Normal vs. Pulmonary stenosis

- Pulmonary valve in normal condition
- Narrowed valve opening and thickened leaflets in pulmonary stenosis

Echo PS_PAIVS_PR SCAI 2014
Echocardiography - Focus

- RVOT region(s) of stenosis -
  - Infundibular hypertrophy
- Pulmonary regurgitation
  - Flow reversal in branch pulm arteries
- RV size, thickness, and function
- TV competency
  - TR severity
  - Estimated RV pressure
  - Hepatic vein systolic flow reversal
  - RA size
- LV size and function
RV Outflow Obstruction

- Imaging and Doppler evaluation
  - Dimensions
- Assess for multiple levels of obstruction and max/mean gradient
  - Infundibulum
  - Valve
  - Supravalve
  - Branch PA’s
Anatomy – Pulmonary Valve Stenosis
Pulmonary Valve Stenosis

- Prevalence = 8-12% of all CHD
- Familial recurrence rate = 2 - 4% in 1st degree relatives
- Noonan’s syndrome
  - 50% of patients have form with PS
  - autosomal dominant transmission
  - 25% offspring affected
Pulmonary Valve Stenosis

- Domed – 42%
- Tricuspid - 6%
- Bicuspid - 10%
- Unicommissural - 16%
- Hypoplastic annulus - 6%
- Dysplastic - 20%
Pulmonary Valve Stenosis

- Post-stenotic dilatation
  - Eccentric Flow
  - Not related to PS severity
- May extend into LPA
- May not be present in newborn
- Not usual with dysplastic PV
### Pulmonary Valve Stenosis - Severity

<table>
<thead>
<tr>
<th>Severity</th>
<th>Peak Doppler (mmHg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trivial</td>
<td>&lt;25</td>
</tr>
<tr>
<td>Mild</td>
<td>25-40</td>
</tr>
<tr>
<td>Moderate</td>
<td>40-60</td>
</tr>
<tr>
<td>Severe</td>
<td>&gt;60 (70)</td>
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</tbody>
</table>

Ductal dependent, Good Echo/Cath Correlation, but debate on MIG vs Mean
Pulmonary Valve Stenosis - Severity

- Both MIG and Mean Doppler grad had good correlation with Cath gradient
- Mean Doppler gradient had significantly less bias
  - Mean Doppler bias: -5 mm Hg
  - MIG bias: +21-26 mm Hg

*J Am Soc Echocardiogr 2005;18:1137-1142*
RV Evaluation – 2D Echo vs Cath

Mean Doppler Gradient Correlated Best with Cath

Brown DW et al. JASE 2012;25:383-92
Pulmonary Valve Stenosis - Severity
Pulmonary Valve Stenosis – Natural History

- PS may progress rapidly during infancy
- Mild PS rarely progresses after 2 y.o.
- Valve thickness/mobility does not correlate with progression
Pulmonary Valve Stenosis - Progression

- **CW 40%**
  - 3.3MHz
  - WF 225Hz
  - Max PG: 38 mmHg
  - Mean PG: 22 mmHg

- **CW 65%**
  - 1.8MHz
  - WF 350Hz
  - Max PG: 68 mmHg
  - Mean PG: 38 mmHg
Pulmonary Regurgitation

- Assessment from multiple views
- Width of PR jet
- Color flow for extent of diastolic flow reversal
TOF – Pulmonary Regurgitation

- 52 adult PTS with repaired TOF
- Echo-Doppler compared to CMR
- Pulmonary Regurgitation index (PRi) = Time PR/Time of diastole (B/A)
- PRi correlated to CMR
  - The smaller the ratio, the worse the PR

Li W et al. Am Heart J 2004;147:165-72
Right Ventricle

- Size relative to LV
- RV free wall thickness
- Function
  - RV fractional area $\Delta$
- Volume
  - 3D Echo
  - MRI – Key vol ml/m$^2$
    - $RVEDVi$ – 150-170
    - $RVESVi$ – 80-90
Real Time 3D Echo RV Volume

- Limitations of 3DE RV Vol
  - Visualization of endocardium
  - Field of view
  - Volumes/sec
3DE RV Volume – Freehand 2DE

Dragulescu A et al. JASE 2011;24:1191-98
CMR – RV Dilatation
CMR – PR Fraction

Main Pulmonary Artery Flow

- Stroke Volume = 73.5 ml
- PR Volume = 31.1 ml
- Regurgitation Fraction = 42.1%

Geva Journal of Cardiovascular Magnetic Resonance 2011 13:9
CMR – RV Evaluation

Geva Journal of Cardiovascular Magnetic Resonance 2011 13:9
Good correlation of 3 Point PR scale with CMR

Brown DW et al. JASE 2012;25:383-92
RV Evaluation – 2D Echo vs CMR

- RV Dias Area $\geq 30$ cm$^2$/m$^2$
- CMR RVEDV $\geq 160$ ml/m$^2$

Brown DW et al. JASE 2012;25:383-92
RV Evaluation – 2D Echo vs CMR

- RV Dias Area ≤20 cm²/m²
- CMR RVEDV < 170 ml/m²

Alghamdi MH et al. JASE 2012;25:518-23
Pulmonary Atresia/Intact Ventricular Septum

- 0.045-0.085/1000 live births
- Heterogenous presentation
  - Variable RV size and function
  - Variable TV size and competency
  - TR associated with RV failure
  - Variable PV and MPA morphology
  - Variable coronary anatomy
Echocardiography PA/IVS - Goal

- Identify patients for RV decompression vs shunt alone (?vs transplant)
- Biventricular, 1½ ventricular, vs Fontan physiology
  - RV size/morphology/growth
  - TV size/morphology/growth
- Initial procedure ultimately depends on identifying RV dependent coronary circulation - angiography
Echocardiography - Focus

- PV size and anatomy
- Architecture of pulmonary arteries
- Infundibular anatomy
- RV to coronary artery sinusoids/fistulas
  - Other coronary artery abnormalities - up to 30-50%
- RV size and morphology (tri-, bi-, uni-partite)
- TV size – absolute and relative to MV
- TV competency
  - RV wall thickness
  - PA size
- Source(s) of pulmonary blood flow
  - Usually single left PDA
  - Tortuosity
- ASD
PA/IVS – Pulmonary Valve and Infundibulum
PA/IVS – Right Ventricle and Tricuspid Valve
PA/IVS – Right Ventricle and Tricuspid Valve

- 30 Patients
- No patients with TV Z-score > -2.5 had RVDCC
  - RV decompression
  - Preop Angiogram
- Addition of color Doppler identified all with coronary fistula

PA/IVS – Right Ventricle and Tricuspid Valve

- 36 PTS
- Low TV Z-scores and TV/MV ratio associated with failed biventricular repair
- TV/MV > 0.5 best predictor of successful biventricular repair

Minich L et al. Am J Cardiol 2000;85:1325-28
PA/IVS – Doppler and Coronary Arteries

- Coronary flow pattern
- Abnormal branching
- Ventriculocoronary connections/Sinusoids
PA/IVS – Tricuspid Competency
PA/IVS – PDA
PA/IVS – ASD
PA/IVS – Fetal Evaluation
PA/IVS – Fetal Evaluation

- 27 live-born followed prenatal
- 19 bivent / 8 nonbivent
- TV/MV diam < 0.7
- TV inflow duration (% cardiac cycle) < 31.5%
- RV/LV length < 0.6
- RV sinusoids

Roman KS et al. Am J Cardiol 2007;99:699-703
PA/IVS – Fetal Evaluation

• Fetal echo 28 PTS
• TV z-score <-4 differentiated outcome groups
• TV growth - groups already differentiated by 23 weeks gest age
• RV:LV width and length also predictive

Biventricular vs Single Ventricle Path

- Coronary artery stenoses/RV dependent coronary circulation
- RV size, morphology, and growth potential
  - Absence/muscularization of infundibulum
- TV size and function
Thank You
Tricuspid Valve

- Severity of regurgitation
- RA size
- Maximal velocity
  - Check multiple sites
- Hepatic vein systolic flow reversal