Surgery for Aortic Root Dilatation Following Repair of Congenital Heart Disease

Christian Pizarro, MD
Alfred I. Dupont Hospital for Children
Wilmington, DE . USA
No disclosures
Root aneurysm / CHD

- Increasing recognition during follow up of patients undergoing interventions for conotruncal anomalies
- Surgical indication not well defined due to unknown natural history
  - risk of rupture or dissection
- Multiple challenges
  - Technically complex
  - Exposure
  - Multiple operations
  - Challenging physiology
  - Surgical risk?
Neo-Aortic Root Dilation and Valve Regurgitation Up to 21 Years After Staged Reconstruction for Hypoplastic Left Heart Syndrome

Meryl S. Cohen, MD,* Bradley S. Marino, MD,* Duff B. McElhinney, MD,* Danielle Robbers-Visser, MD,* Wendy van der Woerd, MD,* J. William Gaynor, MD,† Thomas L. Spray, MD,† Gil Wernovsky, MD*

Philadelphia, Pennsylvania

Figure 2. The diameters of the neo-aortic valve annulus (left), root (middle), and sinotubular junction (right) are plotted against body surface area (BSA) and compared with the normal distribution (mean with 95% confidence intervals).
Long-Term Predictors of Aortic Root Dilation and Aortic Regurgitation After Arterial Switch Operation

Marcy L. Schwartz, MD; Kimberlee Gauvreau, ScD; Pedro del Nido, MD; John E. Mayer, MD; Steven D. Colan, MD

Aortic Root Dilatation in Adults with Surgically Repaired Tetralogy of Fallot
A Multicenter Cross-Sectional Study

- Aortic root dilatation prevalence (30% > 4 cm; O/E >1.5 is 6.6%). Asc aorta >4 cm 19%
- Associated with older age at surgery, pulmonary atresia and mod-severe AI
- Mod-severe AI in 3.5% cases
- Histology strikingly similar to Marfan syndrome.
- Several case reports of dissection

Fate of the Aortic Root Late After Ross Operation

Giovanni Battista Luciani, MD; Gianluca Casali, MD; Alessandro Favaro, MD; Maria Antonia Prioli, MD; Luca Barozzi, MD; Francesco Santini, MD; Alessandro Mazzucco, MD

**TABLE 3. Risk Factors for Autograft Dilatation**

<table>
<thead>
<tr>
<th>Cox proportional hazard</th>
<th>Beta</th>
<th>Standard Error</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>-0.07</td>
<td>0.04</td>
<td>0.05</td>
</tr>
<tr>
<td>Preoperative sinus Valsalva diameter</td>
<td>0.24</td>
<td>0.12</td>
<td>0.02</td>
</tr>
<tr>
<td>Root replacement technique</td>
<td>2.80</td>
<td>1.27</td>
<td>0.03</td>
</tr>
<tr>
<td>Pericardial strip buttressing</td>
<td>-2.61</td>
<td>1.33</td>
<td>0.04</td>
</tr>
</tbody>
</table>

**TABLE 4. Risk Factors for Autograft Dysfunction**

<table>
<thead>
<tr>
<th>Cox proportional hazard</th>
<th>Beta</th>
<th>Standard Error</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (female)</td>
<td>3.51</td>
<td>1.14</td>
<td>0.002</td>
</tr>
<tr>
<td>Preoperative Sinus Valsalva diameter</td>
<td>0.34</td>
<td>0.16</td>
<td>0.04</td>
</tr>
<tr>
<td>Follow-up Sinus Valsalva diameter</td>
<td>0.63</td>
<td>0.21</td>
<td>0.003</td>
</tr>
<tr>
<td>Follow-up sinotubular junction diameter</td>
<td>0.77</td>
<td>0.32</td>
<td>0.02</td>
</tr>
</tbody>
</table>

Luciani Circ 2003;108[suppl II]:II-61-II-67
Sections of aorta show accumulation of myxoid material (extracellular ground substance) within the media.

[H&E, original magnification 40X (left) and 100X (right)]

Elastin stain of aorta shows disruption and loss of elastic fibers within the media.

[Elastin stain, original magnification 100X]

Aorta stained with trichrome stain (left) and smooth muscle actin immunohistochemistry (right) shows expanded zones of extracellular ground substance and disruption and loss of smooth muscle cells within the media.

[Trichrome stain (left), SMA (right), original]
Risk factors for Neo Aortic root dilatation

- Pulmonary artery banding
- Presence of a VSD
- Taussing-Bing anomaly
- Coronary transfer technique?
- Disruption of STJ?
- Disruption of vasa vasorum?
Etiology

- Aneurysm panel, MYLK
- Mutations
  - TGFBR2
  - SMAD 3 mutation
  - MYH11
  - MYLK variant
Aortic Aneurysm in CHD

- Redo sternotomy (2-4\textsuperscript{th})
- Aortic arch replacement in some cases
- Fontan physiology (preserve lung function, cannulation, blood utilization)
- Multiple systemic-pulmonary collaterals
- Increased difficulty of valve sparing procedure after Lecompte maneuver
- Consider associated conditions (decreased ventricular function)
Intervention and Timing

• Lack of natural history data
• Do implications about aneurysm size apply?
• Lack of experience with interventions in these patient population (post arterial switch, Fontan)
• Indications are based on extrapolations from Marfan experience and/or associated pathology
Preoperative Imaging

- Echocardiogram
- CT angiogram
- MRI/MRA

- Trend of growth
- Associated valvar issues
- Involvement of adjacent structures
Technical Aspects

- Full root replacement
- Stabilization of the annulus (reimplantation technique/ David V)
- Arch replacement is not mandatory (Fazel et al)
- Need for routine life long surveillance (aorta at risk)
Clinical data

Cohort included 16 patients

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yrs)</td>
<td>13.5 (5-20)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>45.5 (17-103)</td>
</tr>
<tr>
<td>Height (cm)</td>
<td>155 (113-192)</td>
</tr>
<tr>
<td>Asc Ao (cm)</td>
<td>4 (2-5.1)</td>
</tr>
<tr>
<td>Asc Ao z-score</td>
<td>6.4 (5.4-9.5)</td>
</tr>
<tr>
<td>Ao root (cm)</td>
<td>3.8 (2.4-5.5)</td>
</tr>
<tr>
<td>Ao root z-score</td>
<td>6.2 (4.4 - 13)</td>
</tr>
</tbody>
</table>
Diagnosis

- HLHS 5
- TGA 3
- IAA/Coa 2
- BAV/AS/AI 2
Diagnosis

Bicuspid semilunar valve in 8/16 (50%)

- HLHS 5
- TGA 3
- IAA/Coa 2
- BAV/AS/Al 2
## Previous interventions

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan</td>
<td>5</td>
</tr>
<tr>
<td>Arterial switch</td>
<td>3</td>
</tr>
<tr>
<td>IAA / VSD</td>
<td>2</td>
</tr>
<tr>
<td>Coarctation</td>
<td>2</td>
</tr>
<tr>
<td>Aortic valvuloplasty</td>
<td>2</td>
</tr>
<tr>
<td>DORV, TOF, AVR</td>
<td>1</td>
</tr>
</tbody>
</table>
Indications

- Progressive root dilatation: 16 cases (41%)
- Valve regurgitation: 15 cases (39%)
- PA stenosis: 4 cases (10%)
- Chest pain: 4 cases (10%)

Aortic program
Aortic program

Procedures

- VSARR 7
- Bentall 4
- Tailoring 3
- Ross/Tailoring 2
Valve preservation

- Competency
- Mechanism of regurgitation
- Anatomy / Integrity
- Annular dilatation
- Associated lesions
- Ventricular function
Associated procedures

• Arch repair 4
• PA plasty 3
• Konno 2
• MPA replacement 2
• PV replacement 1
• Ventricular aneurysm repair 1
• Cryo Maze/ pacemaker 1
• Tricuspid valvuloplasty 1
### Clinical data

<table>
<thead>
<tr>
<th>Metric</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>AXC (min)</td>
<td>109 (57-183)</td>
</tr>
<tr>
<td>CPB (min)</td>
<td>175 (103-264)</td>
</tr>
<tr>
<td>DHCA use (%)</td>
<td>6/16 (38%)</td>
</tr>
<tr>
<td>DHCA (min)</td>
<td>29.5 (14-45)</td>
</tr>
<tr>
<td>Mech Ventilation (days)</td>
<td>1 (1-11)</td>
</tr>
<tr>
<td>ICU stay (days)</td>
<td>4 (2-15)</td>
</tr>
<tr>
<td>Hospital stay (days)</td>
<td>11 (6-56)</td>
</tr>
</tbody>
</table>

Aortic program
### Clinical data

<table>
<thead>
<tr>
<th>Category</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>AXC (min)</td>
<td>109 (57-183)</td>
</tr>
<tr>
<td>CPB (min)</td>
<td>175 (103-264)</td>
</tr>
<tr>
<td>DHCA use (%)</td>
<td>6/16 (38%)</td>
</tr>
<tr>
<td>DHCA (min)</td>
<td>29.5 (14-45)</td>
</tr>
<tr>
<td>Mech Ventilation</td>
<td>1 (1-11)</td>
</tr>
<tr>
<td>ICU stay (days)</td>
<td>4 (2-15)</td>
</tr>
<tr>
<td>Hospital stay (days)</td>
<td>11 (6-56)</td>
</tr>
</tbody>
</table>

No operative mortality
HLHS

- Single ventricle physiology
- Multiple reoperations
- Myocardial preservation
- Younger patients
- Compression of the Fontan circuit
- Simpler coronary reimplantation
- Sub pulmonary conus
- Preservation of low PVR
  - CPB time
  - Blood products
Transposition of the great arteries

- Posterior location of the aorta
  - Dissection
  - Hemostasis
- Difficult assessment of the valve sparing procedure
- Coronary anatomy (imaging)
- Pulmonary artery reconstruction
- Older patients
<table>
<thead>
<tr>
<th>Complication</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pleural/peric effusions</td>
<td>2</td>
</tr>
<tr>
<td>Post op bleeding</td>
<td>2 (no reop)</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>3</td>
</tr>
<tr>
<td>Neurologic</td>
<td>1 (periph nerve)</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>1</td>
</tr>
<tr>
<td>Pancreatitis</td>
<td>2</td>
</tr>
<tr>
<td>Sternal wound infection</td>
<td>1 (MSSA)</td>
</tr>
<tr>
<td>GI bleed</td>
<td>1</td>
</tr>
</tbody>
</table>
Follow up

<table>
<thead>
<tr>
<th></th>
<th>VSRR 7</th>
<th>Bentall 4</th>
<th>Tailoring/Ross 5</th>
</tr>
</thead>
<tbody>
<tr>
<td>AI &gt; mod</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Reoperation</td>
<td>3 (OHT, AVR, pseudoan)</td>
<td>2 (AVR, OHT)</td>
<td>0</td>
</tr>
<tr>
<td>Late Death</td>
<td>2 (OHT, pseudoan)</td>
<td>1 (OHT)</td>
<td>0</td>
</tr>
</tbody>
</table>

Median follow up of 51.6 months (30.6-78.2)
Summary

• Despite the high complexity, surgical management of aortic dilatation in patients with CHD can achieve excellent functional outcomes.
• Indications for surgery are usually influenced by coexisting issues (valve incompetence, coarctation, pulmonary artery obstruction).
• Use of valve sparing techniques can effectively restore the aortic or neo-aortic valve competency.
• Durability of these interventions should be monitored over time.
• Genetic information may inform the management of this entity.