Valve Sparing Root Replacement in Congenital Heart Disease

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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
  - Multiple redo
  - Challenging physiology
  - Surgical risk?

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Aortic program
Neo-Aortic Root Dilation and Valve Regurgitation Up to 21 Years After Staged Reconstruction for Hypoplastic Left Heart Syndrome

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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).

JACC 2003;42:533-40
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

Mechanism

Developmental defects in cellular-cellular interactions between SMC and adhesion molecules may provide unique molecular pathways for BAV and TAV associated TAA.

differential expression of cell adhesion and ECM structural proteins between BAV and TAV

Congenital Skills

AATS 2012

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Preoperative Imaging

- Echocardiogram
- CT angiogram
- MRI/MRA

Trend of growth
Associated valvar issues
Involvement of adjacent structures
Case selection
Valve selection

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

- HLHS 4
- BAV/Coa 6
- TGA 3
- DORV 1
- TOF 1

Aortic program
Procedures

- VSARR: 7
- Bentall: 4
- Tailoring: 2
- Ross/Tailoring: 2

Aortic program
Associated procedures

- Arch repair 4
- LPA plasty 2
- Konno 2
- MPA replacement 1
- PV replacement 1
- Ventricular aneurysm repair 1
### Clinical data

<table>
<thead>
<tr>
<th></th>
<th>Value</th>
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<tbody>
<tr>
<td>Age (yrs)</td>
<td>11 (5-18)</td>
</tr>
<tr>
<td>Ao root z-score</td>
<td>6.2 (4.4 - 13)</td>
</tr>
<tr>
<td>Asc Ao z-score</td>
<td>6.4 (5.4 – 9.5)</td>
</tr>
<tr>
<td>AXC (min)</td>
<td>105 (62-172)</td>
</tr>
<tr>
<td>CPB (min)</td>
<td>194 (103-274)</td>
</tr>
<tr>
<td>ICU stay (days)</td>
<td>2 (1 – 4)</td>
</tr>
<tr>
<td>Hospital stay (days)</td>
<td>7 (5 – 12)</td>
</tr>
</tbody>
</table>

No operative mortality  
At a median follow up of 4.6 (0.7- 5.7) yrs  
patients remain free of aortic insufficiency
Perfusion

• Moderate hypothermia unless DHCA used
• DHCA in 5/15 (median 22 min [14-45])
• Pulmonary venous cannulation in Fontan (maintain ventilation)
• Crystalloid cardioplegia
• Cell saver
• Fresh whole blood (median 400 cc[300-550])
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
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 magnifications 100X]
Summary

• Aortic root dilatation after repair of CHD is becoming increasingly recognized.
• Preliminary data suggest a common histological lesion
• In the absence of natural history data and understanding of the surgical risks the indication and timing of surgery remains difficult.
• Indications for surgery are usually influenced by coexisting issues (valve incompetence, coarctation, pulmonary artery obstruction).
• Despite the high complexity of these repairs surgical and functional outcomes are excellent.
Summary

• Developmental defects in cellular-cellular interactions between SMC and adhesion molecules may provide unique molecular pathways for BAV and TAV associated TAA.
• Is the histological lesion similar to aneurysmal dx in the adult?
• Is there a common mechanism? Or just the common expression?
• Can we anticipate who will develop aneurysmal disease following repair of CHD?
• Is this related to the presence of a BV?
• Does the mechanism vary depending on the presence of bicuspid semilunar valve?