Advances in the Evaluation and Management of Valvular Heart Disease in Children

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OptumHealth Webinar
Jan 24, 2013
Overview – Valvular Heart Disease

• Epidemiology of VHD in Children
• Evaluation and Indications for Intervention
• Advances in Evaluation and Management
  – Standardizing Evaluation by Imaging
  – Tetralogy of Fallot / Pulmonary Valve
  – Mitral Valve repair / replacement
  – Concept of “expandable” prosthetic valve
• Future Directions
Cardiac Anatomy 101
Epidemiology and Presentation

• Definition of valve disease
  – 2% of normal population has bicuspid aortic valve
  – Of 8 in 1000 children with CHD, 1/3 have pathology of valve that requires management
• At BCH, approximately 200-300 valvular operations per year
• Physical exam - Murmur
• Valve disease can lead to
  – Symptoms of Heart failure
  – Ventricular dysfunction
Evaluation - Imaging

- Echocardiography is mainstay of diagnosis
- Frequency of Echo follow up depends upon diagnosis
- Detects worsening of valve function
- Ventricle dilation
- Echo changes may precede clinical deterioration
Standardized Clinical Assessment And Management Plans (SCAMPS)

### Table 2. Summary of ASO SCAMP Testing Recommendations

<table>
<thead>
<tr>
<th>Clinic Visit</th>
<th>Outpatient Visit</th>
<th>CXR</th>
<th>Lipid Profile</th>
<th>ECHO</th>
<th>CMR</th>
<th>Stress ECHO or Nuclear Perfusion Scan†</th>
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</thead>
<tbody>
<tr>
<td>1–3 weeks postrepair (0–4 weeks)</td>
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<tr>
<td>6–12 weeks postrepair (4–12 weeks)</td>
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<td>6 months postrepair (4–9 months)</td>
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<tr>
<td>12 months postrepair (9–18 months)</td>
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<td>x</td>
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<tr>
<td>2 years of age (18 months–2 years)</td>
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<td>4 years of age (3–4 years)</td>
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<tr>
<td>6 years of age (5–6 years)</td>
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<tr>
<td>8 years of age (7–8 years)</td>
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<tr>
<td>10 years of age (9–10 years old)</td>
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<tr>
<td>12 years of age (11–12 years old)</td>
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<tr>
<td>14 years of age (13–14 years old)</td>
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<tr>
<td>16 years of age (15–16 years old)</td>
<td>x</td>
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<tr>
<td>18 years of age (17–18 years old)</td>
<td>x</td>
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</tbody>
</table>

x Indicates testing is recommended at that visit.
†Stress echocardiogram (ECHO) recommended. Nuclear perfusion scan is recommended in place of stress echocardiogram if acoustic windows are poor.
ASO, arterial switch operation; SCAMP, Standardized Clinical Assessment and Management Plan; CMR, cardiac MRI; CXR, chest X-ray.
Standardized Clinical Assessment And Management Plans (SCAMPS)

- Goal is to rationalize resource utilization based upon data
- Reduce frequency of unnecessary studies
- Reduce interpractioner variability
- Outcomes can feedback into SCAMP revision
- Can study variations in practice and revise SCAMP or fill gaps with education

<table>
<thead>
<tr>
<th>SCAMP</th>
<th>Date of SCAMP Initiation</th>
<th>Number of Patients</th>
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<tbody>
<tr>
<td>Arterial switch operation</td>
<td>03/22/09</td>
<td>149</td>
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<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>04/05/09</td>
<td>124</td>
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<tr>
<td>Dilated aorta</td>
<td>08/30/09</td>
<td>274</td>
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<tr>
<td>Aortic stenosis</td>
<td>09/05/09</td>
<td>37</td>
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<tr>
<td>Aortic regurgitation</td>
<td>10/18/09</td>
<td>32</td>
</tr>
<tr>
<td>Interstage single ventricle</td>
<td>11/22/09</td>
<td>6</td>
</tr>
<tr>
<td>HLHS with MS/AA</td>
<td>In development</td>
<td>—</td>
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<tr>
<td>Chest pain</td>
<td>In development</td>
<td>—</td>
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<tr>
<td>Small PDA</td>
<td>In development</td>
<td>—</td>
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<tr>
<td>Syncope</td>
<td>In development</td>
<td>—</td>
</tr>
<tr>
<td>Dyslipidemia</td>
<td>In development</td>
<td>—</td>
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</tbody>
</table>

*Enrollment numbers as of 1/20/10.
SCAMPs, Standardized Clinical Assessment and Management Plans; CHB, Children’s Hospital Boston; HLHS, hypoplastic left heart syndrome; MS/AA, mitral stenosis aortic atresia; PDA, patent ductus arteriosus.
Pulmonary Valve

Major Diagnoses

- Critical Pulmonary Stenosis
- Pulmonary Atresia
- Tetralogy of Fallot
Tetralogy of Fallot

Traditional Repair
Long Term – Valve Leakage
Sequelae of Valve Leakage
Indications for Pulmonary Valve Replacement

RV end-diastolic volume index ≥160 mL/m² (Z score >5)
RV end-systolic volume index ≥70 mL/m²
LV end-diastolic volume index ≤65 mL/m²
RV ejection fraction ≤45%
RVOT aneurysm

Clinical criteria: exercise intolerance, symptoms and signs of heart failure, cardiac medications, syncope, sustained ventricular tachycardia.
Valve Replacement

- Options for valve replacement - Bioprosthetic vs. mechanical
- Durability 8-10 years
- Multiple reoperations over lifetime
- Mechanical Valves rarely used – risk of clotting
Advances in TOF - Transcatheter Valve in RVOT

- Can only perform if there is landing zone
  - Previous bioprosthetic valve
  - RVOT stenosis
Advance in Tetralogy – Valve Sparing Repair

- Preserve valve, but enlarge above and below valve
- Balloon dilation of valve
- Valve growth over time
- Potentially avoid any reoperations in future
- 32% compared to 97% regurgitation
Mitral Valve

Major Diagnoses

- Congenital Mitral Stenosis
- Atrioventricular Canal Defect
Cardiac Physiology 101
Congenital Mitral Stenosis
Advance – Mitral Valve Repair

Techniques for Mitral valve repair have evolved over past 10 years.

- Papillary muscle thinning
- Leaflet augmentation
- 90% of valves can be repaired
- Still 50% require repeat operation in 2-3 years
Mitral Valve Replacement

- If valve is irreparable, replacement is an option
- Smallest sized valve is 15 mm
- Most infants have annulus size 8-12 mm
- Oversizing leads to poor left atrial compliance
- Valve stenosis with patient growth
Advance - Mitral Valve Replacement with “expandable valve”

- Melody Valve maintains function over several diameters
- Range of diameter 9-18 mm
- Could implant in infant
- As child grows, could dilate valve in cath lab
- Avoids reoperation if it works
Advantages of Mitral Valve Replacement with “expandable valve”

- Avoid tolerance of residual disease (often associated with RV hypertension)
- Extremely thin wall (Better indexed orifice area)
- Avoid oversizing of valve
- Covered housing prevents pannus ingrowth onto valve leaflets
Advance - Mitral Valve Replacement with “expandable valve”

- Implanted in 9 patients since 2010
- Short term function of valve excellent
- Dilation of valve 3-6 months later (somatic growth)
- 1 patient developed endocarditis – valve explanted
- All others have good valve function at 1 year follow up
Aortic Valve

Major Diagnoses

- Congenital Aortic Stenosis
- Bicuspid aortic valve
Aortic Stenosis – Current Therapy

- Balloon dilation of aortic valve has become mainstay of therapy.
- Surgical commissurotomy rarely necessary - if balloon fails
- Risk – Aortic regurgitation
- Many children avoid operation until adolescence or adulthood
Aortic Stenosis – Surgical Therapy

• Valve regurgitation leads to high left atrial pressures
• Repair involves augmentation of leaflets, debridement of leaflets, tricuspidization of bicuspid valve
• 70% freedom from reoperation at 5 years
• Materials for valve repair suboptimal - calcification
Summary

• Standardized management algorithms rationalize follow up and reduces resource utilization
• Valve sparing repair of Tetralogy of Fallot may improve long term results and reduce reoperation
• Advances in mitral valve repair improve outcomes
• Expandable mitral valve replacement may reduce reoperations
• Future focus on improving materials for valve repair
Contact Us

For more information, please contact me at sitaram.emani@cardio.chboston.org

For consultation or to refer a patient to Boston Children’s Hospital, please contact the Heart Center at 617-355-4278 bostonchildrens.org/heart