Current Strategies for Surgical Treatment of Adult Congenital Heart Disease

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Objectives

- Review indications for surgery in patients with adult congenital heart disease
- Discuss current surgical volumes at IMC
- Describe strategies for ongoing patient management
Case Example

- 40 year old farmer from Blackfoot Idaho
- Repair of tetralogy of Fallot at age 3
- Developed mild exercise intolerance last summer
- Echocardiography demonstrated severe pulmonary insufficiency, severe dilation of the right ventricle and reduced RVEF of 32%
- MRI confirmed above findings
Case Example
Case Example

- Underwent successful redo sternotomy and replacement of pulmonary valve with 29 mm tissue bioprosthesis
- Smooth postoperative recovery
- Chest x-ray at 3 weeks demonstrated reduced right ventricular size
- Echo at 2 months demonstrated improved RV function
Case Example
Case Example
Background

- Milestones in surgery for congenital heart disease
  - PDA ligation, Robert Gross, 1938
  - Extracardiac shunt, Alfred Blalock, 1944
  - ASD closure, John Gibbon, 1953
  - VSD/TOF repairs, Walt Lillehei, 1955
  - Atrial switch, William Mustard, 1964
  - Fontan operation, Francis Fontan, 1968
  - HLHS palliation, William Norwood, 1979
The “Blue Baby” Operation
Background

• Early operations (closed heart) were palliative – effective treatment but not curative
• Later operations (open heart) now focus on “corrective” surgery for more durable results
• “Cure” remains an elusive concept
  • Incomplete cardiac development = need for prosthetic material
  • Life-long management
Definitions

• Adult congenital heart disease:
  • Individual is ≥ 18 years old
  • Underlying heart defect is present since birth (not acquired)

• In general, three major groups of patients:
  • Previously diagnosed/treated/repaired defect(s) in infancy/childhood
  • Newly diagnosed/unrecognized defect in adult
  • Deterioration of prior palliation
Examples

- Previously diagnosed/treated/repaired defect(s) in infancy/childhood
  - Relatively predictable clinical course
  - Tetralogy of Fallot, bicuspid aortic valve
- Newly diagnosed/unrecognized defect in adult
  - May present with near end-stage disease
  - Large ASD, aortic dissection
- Deterioration of prior palliation
  - Complex, variable heart failure
  - Single ventricle/Fontan
Scope

- Surgical and medical advances now allow up to 85% of pediatric congenital heart disease patients to survive into adulthood
- Over 1 million adults in US with some form of congenital heart disease
- Care of these individuals can be complex and does not fit into traditional models of pediatric or adult medical care
Building the Bridge

• Critical concept of ensuring continual, uninterrupted medical care:
  • Adult congenital cardiology
  • Adult congenital cardiac surgery
  • Multiple other healthcare providers
  • Psychological, economic, and social issues
• The patients and their significant others often do not understand the diagnosis, previous operations, and long-term implications
Surgical Trends in ACHD

- Ionescu-Ittu et al (Canada, 2010)
  - Review of Quebec congenital heart disease database from 1998-2005
  - Rate of congenital surgical procedures in children remained constant over time
  - Rates of valvular and aortic surgery in adults increased by 42-63% over time
  - Likely due to left-sided lesions requiring reoperation or newly diagnosed adults
Surgical Trends in ACHD

Surgical Trends in ACHD

• Intermountain Medical Center, 2010-2016
  • Began identifying need for specific adult congenital heart care
    • 2010-2013 = 68 operations
  • Formalization of collaborative ACHD program under direction of Dr. Hoskoppal in 2014
  • Steady increase in surgical and interventional cardiology volume
    • 2014-2016 = 198 operations
Surgical Trends in ACHD

Surgical Cases Per Quarter

- 2014 Q2 Q3 Q4 2015 Q2 Q3 Q4 2016 Q2 Q3 Q4

- 0 5 10 15 20 25 30

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Surgical Trends in ACHD

- Indications for surgery in ACHD at IMC
  - Aortic valve disease: AVR, root replacement
  - Pulmonary valve disease: PVR, conduit exchange
  - Mitral valve disease: mitral repair, MVR
  - Aortic disease: coarctation, bicuspid aneurysm
  - Atrial septal defect
  - Prior complex repair: multiple valves, single vent
  - Undiagnosed: heart failure, advanced valvular disease
  - Septal hypertrophy
Surgical Trends in ACHD

• Aortic valve disease
  • Developing a long-term strategy that is individualized for each patient
• No perfect valve
  • Early valve-sparing operations for Marfan
  • Aortic replacement in bicuspid valve
  • Valve-in-valve transcatheter therapies for bioprosthetic degeneration
Surgical Trends in ACHD

- Mitral valve disease
  - Repair is ideal
    - Repair before development of heart failure
    - Complex re-repair after AVSD surgery
    - Possible transcatheter therapies for complex ACHD
Surgical Trends in ACHD

• Pulmonary valve disease
  • Tetralogy of Fallot/congenital pulmonary stenosis
    • Inadequate valve tissue from birth
    • RV-PA conduit outgrowth/failure
    • Right heart failure can be unpredictable and may be irreversible
  • Transcatheter therapies are expanding
Surgical Trends in ACHD

- Heart failure
  - Advanced, complex disease
  - End-stage result of palliative operations
  - Complex reoperations
  - Need for advanced mechanical circulatory support and transplantation
Surgical Trends in ACHD

- Surgical growth areas in ACHD at IMC
  - Septal myectomy
  - Reoperations for aortic valve/root disease
  - Mitral valve repair
  - Undiagnosed/prior complex repairs
  - Aortic disease
  - Transcatheter valve-in-valve
Surgical Trends in ACHD

Septal Myectomies Per Quarter

- 2014 Q2 Q3 Q4 2015 Q2 Q3 Q4 2016 Q2 Q3 Q4
- 0 5 10 15 20 25

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Surgical Trends in ACHD

- Transcatheter valve-in-valve therapy for ACHD
  - Degenerated aortic bioprosthesis
  - Degenerated right ventricular outflow tract conduit
  - Native pulmonary valve
  - Tricuspid and mitral valve valve disease
Transcatheter Valve Options in ACHD
Surgical Trends in ACHD

Redo AVR/Root

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Valve-in-Valve TAVR
Surgical Trends in ACHD

Combined AVR Procedures

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Surgical Trends in ACHD

- Zomer et al (Netherlands, 2011)
  - Review of >10,000 adult congenital patients from national database (CONCOR)
  - 20% required surgery within 15 years
  - 40% had a reoperation
    - Coarctation, TOF/PS, AVSD, aortic stenosis most common
  - Third and fourth surgeries carried a 2-3X risk of mortality
Surgical Trends in ACHD

Zomer et al, Circulation 2011;124:2195-2201
Surgical Risk Assessment in ACHD

- Giamberti et al (Italy 2009)
  - Retrospective analysis of single institution ACHD patients 2002-2007
  - Mortality 3.6%
  - Major complications in 15%
  - No single factor associated with death, but increasing number of previous operations, CPB time, CHF, lower Hct, and Fontan operation associated with morbidity
Surgical Risk Assessment in ACHD

Surgical Risk Assessment in ACHD

- Holst et al (Mayo 2011)
  - Retrospective analysis of nearly 1000 ACHD patients undergoing multiple reoperations
  - Mortality 3.6%
  - Cardiac injury at sternotomy in 6%
  - Independent factors associated with death included urgent operation, CPB time, lower EF, and single ventricle physiology
### Table 7. Risk Factors for Early Death

<table>
<thead>
<tr>
<th>Variable</th>
<th>Univariate OR (95% CI)</th>
<th>p Value</th>
<th>Multivariate OR (95% CI)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>History of stroke</td>
<td>3.56 (1.56–8.11)</td>
<td>0.003</td>
<td>6.58 (2.40–18.10)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Creatinine &gt; 2 mg/dL</td>
<td>14.90 (5.29–41.96)</td>
<td>&lt;0.001</td>
<td>15.37 (6.14–38.43)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Ejection fraction (per 10)</td>
<td>0.54 (0.41–0.72)</td>
<td>&lt;0.001</td>
<td>1.10 (1.05–1.14)</td>
<td>&lt;0.001</td>
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<tr>
<td>Single ventricle</td>
<td>4.95 (2.22–11.03)</td>
<td>&lt;0.001</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Urgent operation</td>
<td>9.92 (4.30–22.88)</td>
<td>&lt;0.001</td>
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<tr>
<td>Bypass time (per 10 min)</td>
<td>1.14 (1.10–1.18)</td>
<td>&lt;0.001</td>
<td></td>
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<tr>
<td>Circulatory arrest time (per 10 min)</td>
<td>1.54 (1.02–2.33)</td>
<td>0.039</td>
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<tr>
<td>Cross-clamp time (per 10 min)</td>
<td>1.13 (1.07–1.20)</td>
<td>&lt;0.001</td>
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<tr>
<td>Cardiac injury</td>
<td>2.75 (0.92–8.22)</td>
<td>0.071</td>
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<td></td>
</tr>
<tr>
<td>Postoperative transfusion</td>
<td>3.95 (1.63–9.61)</td>
<td>0.002</td>
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<tr>
<td>Sternotomy #3</td>
<td>2.49 (1.20–5.17)</td>
<td>0.015</td>
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<tr>
<td>Sternotomy #4</td>
<td>3.11 (1.09–8.90)</td>
<td>0.009</td>
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</tr>
</tbody>
</table>

CI = confidence interval; OR = odds ratio.

Optimal Surgical Care in ACHD

• Growing patient population
• Wide range of conditions = highly individualized, coordinated, multidisciplinary approach
• Long-term care strategy is critical
  • Reduce number of sternotomies over lifetime
  • Hybrid/new technology interventions
  • Patient and family education
Questions