Surgical Management of Adults with Congenital Heart Disease: Importance of a Congenital and Aortic Partnership

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Disclosures

- None

Advances in Care for Patients with CHD

- Significant medical and surgical advances
  - Improving surgical techniques
  - ICU and anesthetic care
  - Transcatheter interventions
- Survival to adulthood:
  - 15% \(\rightarrow\) 90%

Brida, M et al. Int J Cardiol CHD. 2020
Advances in Care for Patients with CHD

- Increase in adult patients
  - More adults than children starting in 1990s
  - Now more than double
- Many of the patients with complex disease


ACHD Population in United States

<table>
<thead>
<tr>
<th>Category and Age Group</th>
<th>CHD Severity/Race-Ethnicity</th>
<th>Estimated US Prevalence per 1000 (95% Confidence Interval)</th>
<th>Estimated No. of Individuals (95% Confidence Interval)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CHD severity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All ages</td>
<td>Overall</td>
<td>7.85 (7.79–7.92)</td>
<td>2,425,000 (2,425,000–2,444,000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>0.92 (0.90–0.94)</td>
<td>283,000 (277,000–290,000)</td>
</tr>
<tr>
<td>Children</td>
<td>Overall</td>
<td>13.21 (13.03–13.39)</td>
<td>980,000 (966,000–993,000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>4.66 (4.60–4.73)</td>
<td>142,000 (140,000–144,000)</td>
</tr>
<tr>
<td>Adults</td>
<td>Overall</td>
<td>6.16 (6.10–6.22)</td>
<td>1,444,500 (1,431,000–1,459,000)</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
<td>0.68 (0.66–0.70)</td>
<td>160,000 (155,000–165,000)</td>
</tr>
<tr>
<td>Race-ethnicity</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Children</td>
<td>Non-Hispanic white</td>
<td>13.31 (13.12–13.49)</td>
<td>620,000 (612,000–629,000)</td>
</tr>
<tr>
<td></td>
<td>Non-Hispanic black</td>
<td>12.69 (12.50–12.88)</td>
<td>133,000 (131,000–135,000)</td>
</tr>
<tr>
<td></td>
<td>Hispanic</td>
<td>13.26 (13.06–13.45)</td>
<td>227,000 (224,000–230,000)</td>
</tr>
<tr>
<td>Adults</td>
<td>Non-Hispanic white</td>
<td>6.36 (6.29–6.42)</td>
<td>1,104,000 (1,094,000–1,115,000)</td>
</tr>
<tr>
<td></td>
<td>Non-Hispanic black</td>
<td>5.63 (5.56–5.69)</td>
<td>155,000 (153,000–156,000)</td>
</tr>
<tr>
<td></td>
<td>Hispanic</td>
<td>5.58 (5.52–5.65)</td>
<td>186,000 (184,000–188,000)</td>
</tr>
</tbody>
</table>

ACHD Population in United States

• Often compared to tsunami
• Patients lost to follow-up after aging out of pediatric practice
• Not enough cardiologists training in ACHD
  • Poor reimbursement
  • Lack of exposure
• ACHD care limited to larger academic centers

Standardizing Care

• Creation of accredited ACHD centers of excellence
• Extensive requirements
  • Only 51 centers in 26 states
  • Adjacent to large pediatric centers
• Many ACHD clinics that don’t meet all requirements
Comprehensive Care Center Requirements

- Surgical director certified by ABTS in congenital heart surgery
- Minimum of two surgeons
  - 24/7 availability for consultation or emergency surgery
- Expertise in mechanical support and cardiac transplant
- All surgical patients discussed in multidisciplinary format
  - Includes ACHD cardiology, interventional, EP, imaging, anesthesia, ICU
UWMC ACHD Program

- 7 ACHD cardiologists
- 2 ACHD interventional
  - 1 from Children’s
- 3 EP
- 3 HF/Tx cardiologists
- 1 dedicated anesthesiologist
- CTICU
- Two surgeons

What Type of Surgeon?

- Pediatric Congenital Surgeon
  - Primarily at children’s hospital, may come to adult hospital for adult cases
  - Very little enthusiasm in congenital community
  - More interested in neonatal and infant repairs
  - Adult cases not appealing
What Type of Surgeon?

- Adult Surgeon With Congenital Background
  - Common early on (1990s-2000s)
  - More contemporary surgeons less comfortable
    - Less congenital training
    - Less favorable outcomes

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National Practice Patterns for Management of Adult Congenital Heart Disease

Operation By Pediatric Heart Surgeons Decreases In-Hospital Death

Taras Karamlou, MD, Brian S. Diggs, PhD, Thomas Person, MD, Ross M. Ungerleider, MD, MBA, and Karl F. Welke, MD, MS

- National Inpatient Sample
  - 1988-2003
  - ~40,000 ACHD operations
  - Compared “Adult” to “Pediatric”
- Pediatric surgeons >75% case volume
- Major outcome death
  - Minor outcomes LOS, cost

<table>
<thead>
<tr>
<th>Variable</th>
<th>Weighted Frequency±SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>50±0±1±8</td>
</tr>
<tr>
<td>Men, %</td>
<td>65 (0.099±0.015)</td>
</tr>
<tr>
<td>Female, %</td>
<td>22 (0.071±0.006)</td>
</tr>
<tr>
<td>Patient diagnosis, %</td>
<td>155 (0.41±0.089)</td>
</tr>
<tr>
<td>HLHS</td>
<td>199 (0.48±0.097)</td>
</tr>
<tr>
<td>TGA</td>
<td>26 (0.057±0.051)</td>
</tr>
<tr>
<td>D-TGA</td>
<td>105 (0.28±0.051)</td>
</tr>
<tr>
<td>RVOT</td>
<td>90 (0.21±0.051)</td>
</tr>
<tr>
<td>TAVSD</td>
<td>73 (0.18±0.051)</td>
</tr>
<tr>
<td>Tet</td>
<td>101 (0.25±0.019)</td>
</tr>
<tr>
<td>TAPVR</td>
<td>130 (0.33±0.013)</td>
</tr>
<tr>
<td>Coronary of the aorta</td>
<td>162 (0.41±0.024)</td>
</tr>
<tr>
<td>ASD</td>
<td>507 (131±0.026)</td>
</tr>
<tr>
<td>Ostium primum ASD</td>
<td>99 (2.4±0.17)</td>
</tr>
<tr>
<td>Ostium secundum ASD</td>
<td>30 (0.7±0.7)</td>
</tr>
</tbody>
</table>

ACHD Surgical Population

- Evolution over last 30 years
- Introduction of transcatheter options
  - ASD
  - Pulmonary valve
  - Aortic valve
  - Coarctation stenting
- Improvement in ablative techniques

March 2020

- “So...I have a case I was hoping you could help me with...”
  - 26 yo M, hx of d-TGA
  - Six previous sternotomies
  - 3 previous aortic homografts
  - Stent in L pulmonary artery
  - Aortic endocarditis with severe AI

March 2020-Conduct of Operation

- R axillary cannulation
- Redo Sternotomy
- Takedown of LeCompte
- Aortic Root replacement (19mm St. Jude)
- Cabrol reconstruction of coronary arteries
- Reconstruction of LPA with tube graft
- Temporary closure with Cabrol patch
- Many more procedures in 2020
Ideal Surgeon for ACHD Population

2020-2023 Cases (N=225)

- AVR/Aortic
- PVR/RVOT
- Mitral/LAVV
- Tricuspid
- Septal Defect
- Coronary
- Double Root
- Transplant/MCS
- Pacemaker/EP

Ideal Surgeon for ACHD Population

- Congenital anatomy, physiology, operations
  - Right sided lesions, septal defects
- Transplant/MCS
- Advanced aortic surgical and transcatheter techniques
  - Aortic valve repair
- Coronary
- Understanding of transcatheter technology
- Basic understanding of EP/pacemakers

Congenital Surgeon

Aortic Surgeon

? Surgeon (Congenital)
**Referral Pattern-UW**

**Burke Clinic**
- Aortic Valve disease
- Ascending aortic aneurysm
- Aortic arch pathology

**Mauchley Clinic**
- Pulmonary valve/RVOT pathology
- Septal Defects
- Tricuspid valve/Ebstein’s
- Sequelae of AVSD repair
- Anomalous coronaries
- Transplant evaluations

**Combined Cases**
- Ross procedure
- Aortic valve repair
- Double root replacement
- Aortic root/ascending in combination with complex anatomy
- Complex LVOT obstruction
- Unusual coronary anomalies
Case #1

- 25 yo M with DILV, L-TGA, coarctation.
- Surgical palliation with:
  - Norwood procedure (DKS with aortic arch reconstruction, shunt)
  - Bidirectional Glenn shunt
  - Extracardiac fenestrated Fontan
- Followed for ascending aneurysm
  - Now 7.1cm, severe neo-AI, intermittent chest pain, mild decrease in LV function

Case #1-Surgical Approach

- R axillary artery cannulation, L anterior thoracotomy for LV vent, venous cannulation SVC and femoral vein
- Cool to 18°C, X-clamp, ostial cardioplegia
  - Native aortic root separated from DKS
  - Zone 2 arch replacement with branched graft
  - Bentall (On-x 27/29) to neo-aortic root
  - Native aortic root anastomosed to Bentall graft
Case #1-Final Result (MRA)

Case #2

- 46 year old with heterotaxy syndrome, situs inversus with mirror-image dextrocardia, TAPVR, DORV, and partial AVSD.
  - TAPVR, DORV, partial AVSD all repaired as a child
  - Revision of ASD patch and TV annuloplasty as young adult
- Progressive heart failure symptoms related to severe LVOT obstruction and aortic insufficiency
  - VSD patch calcified
  - Long, narrow LVOT
Case #2-surgical approach

- Redo sternotomy, aortic/RA cannulation
- Ostial cardioplegia
- Resection of AV leaflets and inspection of LVOT
- Incision into RV, near complete removal of VSD patch
- Dacron Konno patch to replace VSD patch and enlarge annulus
  - 23 mm St Jude mechanical valve
Case #2-Final Result

32 yo F with Turner’s syndrome
• 2 successful IVF pregnancies
• Intermittent chest pain after delivery of second child
• ST changes on ETT
• Imaging noted intraseptal LM off of R coronary sinus
• Referred for surgery
Case #3

- Normal LM ostium
- No evidence of compression at rest
- Left dominant system

Case #3-Surgical Approach

- Transconal Unroofing as described by Najm et al.
- Reconstruction of RVOT with autologous pericardial patch
Aortic Valve Replacement-Young Patients

- Aortic valve disease common in ACHD population
  - Often present in 20s/30s
- Reluctance to take coumadin
  - Family planning
  - Active lifestyle
- Promise of TAVR
  - More biologic valves
Mechanical or Biologic Prostheses for Aortic-Valve and Mitral-Valve Replacement


30-40% Mortality at 15 Years!
Long-term outcomes after elective isolated mechanical aortic valve replacement in young adults

The Journal of Thoracic and Cardiovascular Surgery • Volume 148, Number 4

- 1997-2006: 469 isolated mAVR <65 years
- Exclusion: concomitant procedures, CAD, reop, emergencies, active endocarditis
- Mean age: 53.2 ± 9.2


Survival Free From Reoperation

At 10 Years, 1 in 5 Patients is Dead or has had a Reoperation!!!

29% mortality at 15 years!

<table>
<thead>
<tr>
<th>Adverse event</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major Hemorrhage</td>
<td>11.8%</td>
</tr>
<tr>
<td>GI/Abdominal Bleed</td>
<td>8.2%</td>
</tr>
<tr>
<td>Intracranial Bleed</td>
<td>4.5%</td>
</tr>
<tr>
<td>Reintervention</td>
<td>4.5%</td>
</tr>
<tr>
<td>Embolism</td>
<td>4.5%</td>
</tr>
<tr>
<td>Endocarditis</td>
<td>2.3%</td>
</tr>
</tbody>
</table>

50% dead/major event at 15 years!
Cost of AVR in Young Patients

- Young and middle aged AVR patients pay a price in life-years or complications regardless of valve type

- *Can we do better in this population?*

The “Living Aortic Valve Complex”

- Better hemodynamics

- Less infection/endocarditis

- No anticoagulation

- *Improved survival???
Ross is the only aortic valve substitute that has been shown to restore patients to [near] normal life expectancy.

UW Ross Program

- First patient in June 2020
- Total of 90 patients to date
- Excellent clinical and valve outcomes
  - 1% mortality
  - 1% stroke
  - 98% freedom from 2+ AI
Outcomes-UW Ross program

- 5.6% reintervention
- 1 take-back for bleeding
- 1 pseudoaneurysm at 6 months
- 2 balloon dilation of homograft

### Table 3. Short- And Long-Term Outcomes after Ross Procedure

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N = 71</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Short-Term</strong></td>
<td></td>
</tr>
<tr>
<td>Tracheostomy</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>New CCVHHD</td>
<td>1 (1.4%)</td>
</tr>
<tr>
<td>Permanent Pacemaker Insertion</td>
<td>5 (7.0%)</td>
</tr>
<tr>
<td>Reoperation</td>
<td>2 (2.8%)</td>
</tr>
<tr>
<td>ICU Transfer</td>
<td>4 (5.6%)</td>
</tr>
<tr>
<td>ICU Length of Stay</td>
<td>1 (&lt;0.1%)</td>
</tr>
<tr>
<td>Atrial Fibrillation Requiring Interven</td>
<td>12 (17%)</td>
</tr>
<tr>
<td>Pneumonia</td>
<td>4 (5.6%)</td>
</tr>
<tr>
<td>Stroke (within 24 hours)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>TIA</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Prolonged asoepes (~48 hours)</td>
<td>5 (7.0%)</td>
</tr>
<tr>
<td>Wound infection</td>
<td>1 (1.4%)</td>
</tr>
<tr>
<td>In-hospital Mortality</td>
<td>0 (0%)</td>
</tr>
<tr>
<td><strong>Long-Term</strong></td>
<td></td>
</tr>
<tr>
<td>Mortality During Follow-Up</td>
<td>1 (1.4%)</td>
</tr>
<tr>
<td>Valve-Related Reintervention</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Any Reintervention</td>
<td>4 (5.6%)</td>
</tr>
<tr>
<td>Pregnancy</td>
<td>5 (2.9%)</td>
</tr>
</tbody>
</table>

Note: N = 71, Mean (SD)

CCVHHD: Continuous veno-venous hemofiltration/hemodialysis
TIA: Transient Ischemic Attack
ICU: Intensive Care Unit
Lessons Learned

• Delicate between rapid programmatic growth and optimal outcomes
• Dedicated and specialized team essential
  • Congenital and aortic partnership
• “Buy-in” from all involved
  • ICU, outpatient → BP control
• Follow-up essential

Aortic Valve Repair

• Reserved for patients with primary AI

• Commonly associated with root aneurysm → VSRR

• Most BAV AI patients can be repaired

• Rapid area of growth and innovation
Repair Techniques

• AI comes from pathology within → annulus, STJ, or leaflets

• Repair techniques address:
  • Annuloplasty
  • STJ ring
  • Leaflet repair/plication

Case #1-AVr

• 21 yo M with severe BAV AI and 4.9 cm root

• David V with BAV repair

• Post → no AI, MG 7 mmHg
Case #2-AVr

• 40s M with root aneurysm, TAV, moderate AI

• Hx of bleeding-concerned about anticoagulation

TAV with eccentric AI jet!!

Elongated NCC

Prolapse of RCC

David V with asymmetric commissural reimplantation and plication of RCC
Final Thoughts

- ACHD population continues to grow and will need surgical and transcatheter intervention
- Surgery on the aortic valve or ascending aorta is common
- The expertise of a surgeon familiar with complex aortic techniques is critical for the success of an ACHD program
- Partnership between a congenital surgeon and aortic surgeon offers the most comprehensive care for this complex population