Fundamentals of Congenital heart surgery in the adult

Hospital Nacional A Posadas

Hospital Universitario Austral

Christian Kreutzer, MD
AATS Cardiovascular Symposium, Sao Paulo December, 2017
STS CHSD Procedures in Adults ≥ 18 Years

Number of Patients

N=1,942
15.4%

N=8
2
0.7%

Repaired Patients Are Not “cured” nor “Fixed”

- False perception of “cure” fostered by the phrase “total correction”

- In reality, there is almost no surgical cure for most CHD

- Nearly all repaired lesions have the potential for residuae and sequelae.
### Epidemiology

#### Survival to 18 yrs of Age with Simple CHD

<table>
<thead>
<tr>
<th>Decade Born with CHD</th>
<th>Percent Survival to 18 Years Old</th>
</tr>
</thead>
<tbody>
<tr>
<td>1940</td>
<td>30%</td>
</tr>
<tr>
<td>1960</td>
<td>65%</td>
</tr>
<tr>
<td>1970</td>
<td>75%</td>
</tr>
<tr>
<td>1980</td>
<td>85%</td>
</tr>
</tbody>
</table>

Warnes et al. J Am Coll Cardiol 2001;37:1170-75
Epidemiology

Survival to 18 yrs of Age with Complex CHD

Decade
Born with CHD

1980
80

1970
50

1960
15
1% D-TGA and 10% of TOF to adulthood

1940
5

Percent Survival to 18 Years Old

Epidemiology

Distribution of Age at Death in Patients With Congenital Heart Disease in 1987 to 1988 and 2004 to 2005

Greatest survival trend occurred in those with more severe forms of CHD

Epidemiology

Adult CHD Patients

- 325,000 (Hoffman 1978)
- 500,000 (Fyler 1980)
- 750,000 (Ferencz 1985)
- 1,000,000 (Kaiser 1980, New England)
- 1,300,000 (Baltimore-DC 1985)

20,000 new patients/yr
5% increase/yr
Epidemiology

Ratio of Pediatric to Adult Patients with CHD

- Pediatric patients
- Adult patients

1965: 70 Pediatric patients, 30 Adult patients
1985: 50 Pediatric patients, 50 Adult patients
2005: 40 Pediatric patients, 60 Adult patients

Williams et al. J am Coll Cardiol 2006;47:701-7
**Adult Congenital Heart Disease (ACHD) complexity**

**SIMPLE – 47%**
- Simple ASD
- Simple Aortic Disease
- Simple Mitral Disease
- Simple PDA
- Mild PS

**COMPLEX – 15%**
- Mitral Atresia, D-TGA, CCTGA, DORV,
- Heterotaxy, Single V, Conduits, Truncus
- Cyanotic, Eisenmenger

**MODERATE – 38%**
- TOF
- SV-Defects
- APV Drainage
- AVC
- Sub PS
- Ao Co
- Ebsteins
- PS
- VSD

**Arrhythmias.**
- Heart failure,
- Pulm. Hypertension,
- 50% will need repeat interventions.

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*Warnes C, et al. JACC 2001*

*Hoffman JACC Vol. 39, No. 12, 2002*

*Warnes JACC Vol. 37, No. 5, 2001*
In-hospital death rates for patients operated on by pediatric heart surgeons were lower than those operated on by non-pediatric heart surgeons (1.9% versus 4.8%, p<0.0001).

“…likely the result of refined processes of care at institutions possessing specialized congenital heart surgeons in addition to the expertise of the surgeons themselves.”
Adult or Pediatric?

• In the 70’s Coronary artery by pass graft revolution. (René Favaloro)
  – Two Subspecialities
    – Pediatric or Congenital Heart Surgery (Pediatric Hospitals)
    – Adult or Aquired Heart disease Surgery
• Pediatric or Congenital heart Surgery 80’s and 90’s
  • Infant heart surgery
  • Neonatal Heart Surgery.
• For adults with complex CHD it’s time to get back together.
• Back together in a HEART TEAM
## Comorbidities

<table>
<thead>
<tr>
<th>Comorbidity</th>
<th>Cases (%) n=9,952</th>
<th>Controls (%) n=29,837</th>
<th>OR (adjusted)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial fibrillation</td>
<td>425 (4.3)</td>
<td>206 (0.7)</td>
<td>7.6 (6.1-9.3)</td>
</tr>
<tr>
<td>Heart failure</td>
<td>257 (2.6)</td>
<td>136 (0.5)</td>
<td>6.7 (5.2-8.5)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>929 (9.3)</td>
<td>2008 (6.7)</td>
<td>1.4 (1.3-1.5)</td>
</tr>
<tr>
<td>CKD</td>
<td>80 (0.8)</td>
<td>61 (0.2)</td>
<td>3.4 (2.3-5.1)</td>
</tr>
<tr>
<td>Diabetes</td>
<td>256 (2.6)</td>
<td>562 (1.9)</td>
<td>1.3 (1.1-1.5)</td>
</tr>
<tr>
<td>Depression</td>
<td>285 (2.9)</td>
<td>767 (2.6)</td>
<td>1.1 (0.9-1.2)</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>241 (2.4)</td>
<td>283 (0.9)</td>
<td>2.4 (1.9-3.0)</td>
</tr>
<tr>
<td>Erythrocytosis</td>
<td>28 (0.3)</td>
<td>3 (0.01)</td>
<td>23.5 (7.1-77.8)</td>
</tr>
<tr>
<td>Stroke/TIA</td>
<td>257 (2.6)</td>
<td>283 (0.9)</td>
<td>2.6 (2.2-3.2)</td>
</tr>
</tbody>
</table>

*Source: Billett J et al. Heart 2008;94:1194-9*
Outcomes and Costs of Cardiac Surgery in Adults with Congenital Heart Disease

Viviane G. Nase¹ - David Faraoos² - Anne Marie Valente² - James A. Dinardo¹

Fig. 1 Hospital costs for cardiac surgery in adults with CHD compared with CABG

Pediatr Cardiol (2017) 38:1359–1364

Rate Ratio (95% CI)
Risk Factors and Early Outcomes of Multiple Reoperations in Adults With Congenital Heart Disease

<table>
<thead>
<tr>
<th>Variable</th>
<th>Univariate 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>
</tr>
<tr>
<td>Creatinine &gt; 2 mg/dL</td>
<td>14.90 (5.29–41.96)</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>
</tr>
<tr>
<td>Single ventricle</td>
<td>4.95 (2.22–11.03)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Urgent operation</td>
<td>9.92 (4.30–22.88)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Bypass time (per 10 min)</td>
<td>1.14 (1.10–1.18)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Circulatory arrest time (per 10 min)</td>
<td>1.54 (1.02–2.33)</td>
<td>0.039</td>
</tr>
<tr>
<td>Cross-clamp time (per 10 min)</td>
<td>1.13 (1.07–1.20)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Cardiac injury</td>
<td>2.75 (0.92–8.22)</td>
<td>0.071</td>
</tr>
<tr>
<td>Postoperative transfusion</td>
<td>3.95 (1.63–9.61)</td>
<td>0.002</td>
</tr>
<tr>
<td>Sternotomy #3</td>
<td>2.49 (1.20–5.17)</td>
<td>0.015</td>
</tr>
<tr>
<td>Sternotomy #4</td>
<td>3.11 (1.09–8.90)</td>
<td>0.009</td>
</tr>
</tbody>
</table>

Surgical Challenges

• Natural Survivor with complex CHD
• Unnatural Survivor. Palliated or "Corrected"
  – Patients present with variable knowledge
  – Hours worth of reading or sternotomy with unknown history
    • Operative reports are impossible to locate
• What do we have behind the sternum? MRI, Cath
  – Space
  – Aorta
  – RV.
  – RA.
Reentry Issues and residual shunts

- Right atrium (Fontan, Ebstein)
- Right ventricle
- Aorta – TGA, TOF, PA-VSD, Ross
- Conduits – Rastelli,
  - TGA
  - Truncus A.
  - PAVSD
Operative issues

Pre sternotomy: Femoral or axillary/Innominate artery arterial preparation/cannulation

Multiple cardiac catheterizations in Childhood = Loss of femoral vessels
Injury @ re entry

- Always plan ahead. Ready for cannulation prior sternotomy.
- Unavoidable. Conduits, homografts, Fused with sternum.
  - Sternotomy under full peripheral CPB.
- High Risk:
  - Prepare your arterial and venous Access.
  - Aorta.
    - not complete the sternotomy, innomniate artery or axilar cannulation. (Better Brain perfusion)
    - Deep hypothermia.
    - Complete sternotomy in Trendelemburg with low flow or DHCA
  - RV or RA: Go rapidly on by pass, cool and try to repair.
    - Beware of residual shunts (Air embolism and stroke)
Most Common procedures.

- **Pulmonary Valve Replacement**
  - TOF and Rastelli procedure (RV to PA conduit)
- **Aortic valve repair/replacement**
  - Aortic Root Dilation ± Coarctation
    - TOF PA.
    - TGA after arterial Switch.
    - Ross procedure.
    - HLHS and other Single Ventricle.
    - BAV ± Coarctation.
- **Mitral Re Do**
  - AV Canal.
- **Ebstein’s Anomaly**
  - Da Silva’s Cone procedure
- **Fontan Kreutzer Revision and Conversion**
Pulmonary valve Replacement.

- **CHD with PS or PA require a RVOT procedure.**
  - Tetralogy of Fallot. (Valvulotomy or TAP)
  - Pulmonary stenosis. (Surg or Perc. Valvuloplasty)
  - D TGA with PS. (Rastelli, Nikaidoh) (RV PA conduit)
  - LTGA with PS (LV or RV PA conduit)
  - Truncus arteriosusus. (RV PA conduit)

- **Nearly all will fail for Regurgitation.**
  - RV Dilation.
  - RV failure and Exercise Intolerance
  - Arrhytmia and sudden death
Pulmonary valve Replacement.

- Low M/M
  - RV rupture at sternotomy. Stroke
  - RV failure.
  - Arrhythmias.

- Adressing other lesions (TR, residual VSD, PA branch stenosis)

  - Meta analysis 141 papers
  - 12 art Homografts outperform xenografts (Not Stat significant).
  - Two articles have suggested that xenografts outperform homografts.
Mitral or left AV valve repair.

• Post AV canal (20 % rate of reoperation)
  – Cleft mitral valve.
  – Retracted leaflets
  – Patch augmentation.
  – Rings, neo Chordae

• Left or Common AV valve in Single Ventricle
  – SV Dilation.
  – SV failure
  – Replacement a valid alternative
Ebstein’s Anomaly

- Distal attachment of the septal & posterior leaflets away from the AV junction
- Atrialized RV.
  - Inlet part of RV above TV attachment;
  - in symptomatic patient tend to be thin-walled and dilated
- Functional RV – apical and infundibular component:
  - Thinner
  - Hypoplastic
  - fewer than normal muscular fibers
  - contain more fibrous tissue
- A combination of RV failure and TR
Ebstein’s Anomaly.

- Effective repair for TR
- improved clinical status.
- better left ventricle filling
- Improved objective exercise capacity.
Ascending Aortic Aneurism in CHD.

- **BAV ± Coarctation of Aorta**
- **Conotruncal anomalies**
  - TOF
  - Truncus
  - DORV
- **TGA post Arterial switch.**
- **HLHS status post Norwood.**
- **Single Ventricle.**
- **AR main indication for surgery (Dissection is rare).**
Aortic Dissection in CHD: Myth or Truth

Aortic dissection in hospitalized children and young adults: a multiinstitutional Study

- Pediatric health Information system database 2004/2011. 7 yrs 30 mill admissions
- 124 dis. in 110 pts. Male 69 % mean age 12,9 años.
- Diagnosis:
  - CHD (38%)
  - Trauma (24%)
  - CTD (16%)
  - Hypertension (HTN) (8%).

*Cong heart Dis. 2014 Jan-Feb;9(1):54-62.*
A little “Sample”

- 33 y old TOF PA, MAPCAS. 43 Kg NYHA class IV
- Status post Right Blalock Taussig Shunt.
- Left PA stenosis. Large collateral supplying L lung Up Lobe.
- Ascending aorta of 10 cm. AR.
- Art Sat of 71% and Hct: 69 %.
- Complete “Repair”
  - Midline unifocalization, LPA plasty
  - VSD Closure and RV PA 20 mm aortic homograft (Rastelli procedure)
  - Bentall (St Jude Valved Conduit)
  - Ao CX 154 ′, CPB 250
A little “Sample”

- 2 full teams
  - One pediatric
  - One adult
- 11 hs of OR time.
- 4 hs of hemostasis.
- ICUS: 10 days
- LOS: 27 days-
- Works full time as an actor/singer in a musical play.
- Enjoys life.
Conclusion

- Complex patient population
- Reoperation always a risk factor.
- Be prepared and anticipate events
  - Previous plan.
  - Checklist
  - Medical and surgical history.
- Experienced center
  - Experienced pediatric cardiac surgeon
  - Experienced Adult Cardiac surgeon in reoperations and aortic and valve surgery
- Heart team.
Pulmonary valve Replacement.

# Early results

## Risk Factors and Early Outcomes of Multiple Reoperations in Adults With Congenital Heart Disease

<table>
<thead>
<tr>
<th>Sternotomy #</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5+</th>
<th>All</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>630</td>
<td>298</td>
<td>78</td>
<td>34</td>
<td>984</td>
</tr>
<tr>
<td>Mortality (%)</td>
<td>2.3</td>
<td>5.6</td>
<td>6.9</td>
<td>0</td>
<td>(3.6%)</td>
</tr>
<tr>
<td>Resp. failure (%)</td>
<td>5</td>
<td>6</td>
<td>7</td>
<td>16</td>
<td>57 (6%)</td>
</tr>
<tr>
<td>Pacemaker (%)</td>
<td>4</td>
<td>5</td>
<td>4</td>
<td>0</td>
<td>38 (4%)</td>
</tr>
<tr>
<td>Stroke (%)</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>12 (1%)</td>
</tr>
<tr>
<td>Renal failure (%)</td>
<td>3</td>
<td>4</td>
<td>6</td>
<td>3</td>
<td>31 (3%)</td>
</tr>
<tr>
<td>Sternal infect (%)</td>
<td>2</td>
<td>1</td>
<td>7</td>
<td>3</td>
<td>19 (2%)</td>
</tr>
</tbody>
</table>
