The Long View after Congenital Heart Surgery

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Director of Clinical Research
Shaping of a new field: Pediatric Cardiology / Surgery

**Concepts**

**Tools**

**Pioneers**
The ductus arteriosus is ligated and divided (Robert Gross, Boston 1938)

SURGICAL LIGATION OF A PATENT DUCTUS ARTERIOSUS: REPORT OF FIRST SUCCESSFUL CASE
R. E. Gross and J. P. Hubbard

The continued patency of a ductus arteriosus for more than the first few years of life has long been known to be a potential source of danger to a patient for two reasons: First, the additional work of the left ventricle in maintaining the peripheral blood pressure in the presence of a large arteriovenous communication may lead eventually to cardiac decompensation of severe degree. Second, the presence of a patent ductus arteriosus makes the patient peculiarly subject to fatal bacterial endocarditis. While it is true that some persons have been known to live to old age with a patent ductus of Boselli, statistics have shown that the majority die relatively young because of complications arising from this congenital abnormality. Dr. Made Aubert presented a series of eight papers which came to autopsy in which it was shown that the patient had had a patent ductus arteriosus without any other cardiovascular abnormality. Of these patients, approximately one fourth died of bacterial endocarditis of the pulmonary artery and an additional one half died of slow or rapid cardiac decompensation. The average age of death of patients in this series was 24 years.

The complications arising from the persistence of a patent ductus arteriosus would seem to make surgical ligation of this abnormal vessel a rational procedure, if such a procedure could be completed with promise of a low operative mortality. Dramatic results have previously been obtained in persons with cardiac enlargement and decompensation resulting from a peripheral arteriovenous aneurysm when the short-circuited vessels have been ligated or excised. On similar theoretical grounds, future cardiac embarrassment should be averted if a shunt between the aorta and the pulmonary artery could be removed. It would also seem plausible to expect that the shutting off of the abnormal stream of blood passing into the pulmonary artery would lessen the formation of the thickened subendocardial plaques within the pulmonary artery, which are so likely to be the seat of later bacterial infection. The surgical approach to the aortic arch and pulmonary trunk having been studied previously in animal experimentation, it seemed within reason that a patent ductus could be adequately exposed in man and possibly ligated without undue danger. It was therefore decided to undertake the operation in a child who presented the classic signs of a patent ductus arteriosus. At the age of 7 years she already had cardiac hypertrophy, which developed presumably from the embarrassment resulting from the anomalous communication. It was to be expected, therefore, that she would have increasing severe disability in the future, aside from the danger of having bacterial endocarditis develop.
A “ductus” is created: the Blalock-Taussing shunt (Baltimore, 1944)
Direct vision intracardiac correction of Congenital Heart Defects (CHD)

Controlled cross-circulation (Lillehei, Minneapolis 1955)
Fontan and Norwood:
Staged palliation for Single Ventricle lesions

Fontan, 1971
Norwood, 1981
Outcomes after operations for CHD – Pediatric Cardiac Care Consortium (PCCC)

- US-based registry of cardiac interventions
  - Includes 47 US centers
  - Clinical data collected 1982 – 2011

- Linkage to long-term outcome databases through December 31, 2014 for patients entered before April 15, 2003
  - National Death Index (NDI)
  - Organ Procurement and Transplantation Network (OPTN)
Crude and risk-adjusted perioperative mortality rate decreases over time

*J Vinocur et al: Pediatr Cardiology 2013; 34:1226-1236*
Mortality improved over time in all risk categories except category 1

As mortality of children with CHD drops, the number of adult survivors with CHD increase.

Number of adults living with CHD surpassed the number of infants and children with CHD.

Mortality Before Age 18
Long-term Outcomes of CHD

**Mild CHD**
- PDA
- ASD (secundum)
- VSD (isolated)

**Moderate CHD**
- ASD (primum)
- TAPVR and PAPVR
- CAVC or PAVC
- Ebstein’s anomaly
- PS (moderate or severe)
- Sub- or supra-AS
- VSD (complex)
- CoA

**Severe CHD (2-Ventricles)**
- Double-outlet ventricle
- Pulmonary atresia
- d-TGA
- TAC
- L-TGA
- All forms of cyanotic heart disease
- Other complex CHD

**Severe CHD (Single Ventricle)**
CHD are at increased risk for premature death across all groups

Long-term Outcomes of CHD

A. CHD patients surgically treated in the Pediatric Cardiac Care Consortium

- Mild (N=12,594): 99%
- Moderate (N=14,020): 97%
- Severe 2V (N=5,410): 86%
- Single Ventricle (N=3,434): 71%

B. Conditional survival after discharge

- Mild
- Moderate
- Severe 2V
- Single Ventricle
- Matched US Population

C. Cumulative in-hospital and post-discharge survival at 25 years

- Mild: 97%
- Moderate: 93%
- Severe 2V: 86%
- Single Ventricle: 65%
- Matched US Population: 89%

Long-term outcomes improve over time for selected lesions (Atrioventricular canal, Transposition of the great arteries and Single Ventricle)
Years of life lost due to CHD

**Males**

- **Mild**
- **Moderate**
- **Severe 2V**
- **SV**

**Females**

- **Mild**
- **Moderate**
- **Severe 2V**
- **SV**

- Contribution from in-hospital deaths
- Contribution from post-discharge deaths
Late causes of death in patients operated for CHD

- Congenital Heart Disease: 56.0%
- Diseases of the Circulatory System: 11.1%
- Other Congenital Malformation, Deformation, or Chromosomal Abnormality: 4.7%
- Diseases of the Respiratory System: 3.6%
- Infections and Parasitic Diseases: 3.4%
- Neoplasms: 2.1%
- External Causes of Injury and Poisoning: 8.2%
- Other: 11.0%
Causes of death over time
Late causes of death in patients operated for CHD

<table>
<thead>
<tr>
<th>Disease Classification</th>
<th>CHD Caused Death</th>
<th>CHD Associated Death</th>
<th>CVD (Non-CHD) Associated Death</th>
<th>Other Death (Non-CHD, Non-CVD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>57</td>
<td>29</td>
<td>60</td>
<td>161</td>
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<tr>
<td>Moderate</td>
<td>267</td>
<td>69</td>
<td>115</td>
<td>133</td>
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<tr>
<td>Severe 2V</td>
<td>398</td>
<td>74</td>
<td>64</td>
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<td>Severe 1V</td>
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<td>110</td>
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<td>L-R Shunt</td>
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<td>81</td>
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<td>LHOL</td>
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<td>Miscellaneous</td>
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<td>60</td>
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<td>Complex Lesions</td>
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<td>49</td>
<td>40</td>
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<tr>
<td>Overall</td>
<td>1415</td>
<td>307</td>
<td>381</td>
<td>424</td>
</tr>
</tbody>
</table>
Conclusions

• Outcomes for CHD have steadily improved the last 30 years
• Survivors with repaired or palliated congenital heart disease have a significantly higher mortality ratio than the general population
• Most deaths are related to the CHD or cardiovascular conditions, although risk for other causes of death is elevated as well
• External causes of death is the only cause of death from which patients with CHD are protected
• Additional efforts are needed to improve the outcomes of patients with CHD to reach the full life potential of the normal population.
Other imPOSSIBLE dreams of the past…

First flight’s length (105 ft for 3.5”) was shorter than the wingspan of a Boeing 747 (211 ft 5”)!

When Jules Verne was suggesting that one day man will go to the moon, people thought this was an insane idea.

Reaction to Graham Bell’s invention of telephone: This device is impractical and unlikely to be of use to anyone.
"What mankind can dream research and technology can achieve"

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