Complex congenital heart disease

From babies to adults:
A growing problem

Otto H. Teixeira, MD
Prevalence of adult cardiovascular disease in USA: 1 in 3 (in million) (Circulation 2006;113:e85-e151)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Cases (in million)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>65</td>
</tr>
<tr>
<td>Coronary</td>
<td>13.2</td>
</tr>
<tr>
<td>Stroke</td>
<td>5.5</td>
</tr>
<tr>
<td>Heart failure</td>
<td>5</td>
</tr>
<tr>
<td>Congenital *</td>
<td>1</td>
</tr>
<tr>
<td>TOTAL</td>
<td>71.3</td>
</tr>
</tbody>
</table>

*USA 2010
Circulation.
2014;130:749-50
Childhood Diseases:

*Dramatic survival over the last 50 years*

- Cancer – 80 % survival to adulthood
- Cystic fibrosis – 40 % are adults
- Greatest advances: complex diseases:
- 85 % of babies with CHD survive
Survival

- *BirthYear* ([JACC 2001;37:1170])

![Bar chart showing survival in different years and birth years: 1940-1959, 1960-1979, 1980-1989. The chart includes bars for Simple, Moderate, and Complex categories.](image-url)
The lifetime prevalence of congenital heart disease in children and adults in Quebec, Canada, in 2010. 95%CI indicates 95% credible interval.

In 2010, the prevalence was 13.11, 95%CI = (12.43, 13.81) per 1000 children, and 6.12, 95% CI = (5.69, 6.57) per 1000 adults

Change in congenital heart disease prevalence of children and adults in Quebec, Canada, from 2000, 2005, and 2010 for patients with severe congenital heart disease.

The numbers and proportions of adults and children in Quebec, Canada, with all (A) and severe (B) congenital heart disease over time in 2000, 2005, and 2010.

Simple Congenital Heart Disease

- ASD
- VSD
- PDA
- PVS
- AVS, MS
- Bic AO
Moderate Complexity

- Tetralogy of Fallot
- Coarc
- AVSD
- TAPVR, PAPVR
- Ebstein
Complex Congenital Heart Disease

- Transposition of the Great Arteries
- Single Ventricle Physiology
  - Truncus
  - Cyanotic: all forms
Common issues

- Exercise intolerance and heart failure
- Arrhythmia
- Reoperations
- Pregnancy
- Employment and other QOL issues
- Life expectancy
Abnormal exercise test and CHF by age in years

(Am J Cardiol 2006;97:1238)
Arrhythmias (Circ 2007;115:534)

*Tetralogy of Fallot*

- SVT 30 %
- VT 10 %
- AVB/SAN dysf 5 %
Fontan & TGA S/P atrial switch

- SVT 50%
- SAN dysf: increased with age
Dance partners
Case study: TGA

- 9 hour-old BBB with cyanosis and metabolic acidosis not responding to oxygen
- PGE1 infusion started, baby promptly improved
Case study: TGA

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- PGE1 infusion started, baby promptly improved
Case study # 2: TGA

- 19 yo female admitted in severe CHF, sepsis-like picture.
- Hx of Senning procedure
- Apparently well till 1 –2 wks prior admission
- Adult cardiologist consulted, referred pt to pediatric cardiologist (*smart guy!*)
The Problem: 1.5 million and counting

- High maintenance
- Not “cured”
- Potential for *sequelae* even with simple defects
- 50% may have CHF, arrhythmia, or surgery
- Life long F/Up (JACC 2005: 46:1)