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>Disease</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>65</td>
</tr>
<tr>
<td>Coronary</td>
<td>15.2</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* USA 2014</td>
<td>71.3</td>
</tr>
<tr>
<td>TOTAL* USA 2020</td>
<td>71.5</td>
</tr>
</tbody>
</table>

*Data from Circulation 2006;113; e85-e151
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**

**Birth Year (JACC 2001;37:1179)**

The lifetime prevalence of congenital heart disease in children and adults in Quebec, Canada, in 2010. 95%CI indicates 95% credible interval.

Adapted from JACC. Circulation. 2014;130:107-116.
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

Arrhythmias

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

- 9 hour-old BBB with cyanosis and metabolic acidosis not responding to oxygen
- 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)