Sudden Death in Congenital Heart Disease:

What do epidemiologic studies tell us?

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Disclosures

• No financial conflicts
• Excessive fun dancing with Medtronic employee last night though
Case study

- 22 yo medical student with repaired ASD, pacemaker for advanced 2\textsuperscript{nd} degree AV block
- Family History: Father with heart block, 2 sisters with ASDs, cousin with tricuspid atresia
  - Sudden death in father after ENT surgery
Case study

• Patient experienced sudden death
  – Autopsy showed no obvious cause
  – Pacemaker interrogation unrevealing, working properly, had been checked 2 months before
• NKx2.5 mutation found in affected family
• 3 more family members died suddenly

Lesion: epidemiology informs our practice in part, Genetics in part
Complexities of adult CHD that impact survival

• Initial anatomy and genetic make-up
  – eg ToF variants, NKx2.5

• Surgical history, quality of repair
  – eg scars that cause reentry tachycardia

• Late hemodynamic and arrhythmic effects
  – eg poor valve function resulting in ventricular dilation
1st big population-based study of survival after CHD surgery

- 41 unexpected late deaths
  - Almost all in tetralogy, transposition, aortic stenosis, coarctation
  - Event rate 1/454 pt-years in these 4 dx
  - Event rate for all others 1/7174 pt-years

Silka, JACC 1998;32:245-51
Sudden death clustered

Hatched: arrhythmia, Open: circulatory, Dark: CHF
Kaplan-Meier survival curves

Cyanotic defects

Left sided obstructive lesions
Late Causes of Death After Pediatric Cardiac Surgery: A 60-Year Population-Based Study

- Survey of all late deaths following CHD surgery under 15 yo in Finland 1953-2009
- Total population 10,964 patients
  - All-cause late mortality 10%, cardiac 6.6%
  - Sudden death in 1.5%
- 40 yr freedom from SCD 99% for simple defects, 91% for severe defects

Raissadati et al. JACC 2016
Same trends as Silka paper over the duration of the study

* Includes untreated aortic stenosis
Marked decrease in SCD in later cohort

<table>
<thead>
<tr>
<th>Defect</th>
<th>Cardiovascular Death Incidence/1,000 PY</th>
<th>Heart Failure Death Incidence/1,000 PY</th>
<th>Sudden Death Incidence/1,000 PY</th>
<th>Post-Reoperative Early Death Incidence/1,000 PY</th>
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</thead>
<tbody>
<tr>
<td>PDA</td>
<td>0.07</td>
<td>0</td>
<td>0.31</td>
<td>0</td>
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<tr>
<td>ASD</td>
<td>0.08</td>
<td>0</td>
<td>0.15</td>
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<td>COA</td>
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<td>0.58</td>
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<tr>
<td>VSD</td>
<td>0.33</td>
<td>0.08*</td>
<td>1.42</td>
<td>0.84*</td>
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<td>TOF</td>
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<td>0.78</td>
<td>1.94</td>
<td>0.78</td>
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<tr>
<td>TGA</td>
<td>1.28</td>
<td>0.95</td>
<td>4.81</td>
<td>2.21†</td>
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<td>UVH</td>
<td>3.50</td>
<td>3.47</td>
<td>11.51</td>
<td>9.26</td>
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<tr>
<td>Misc</td>
<td>0.62</td>
<td>0.83</td>
<td>3.10</td>
<td>2.95†</td>
</tr>
</tbody>
</table>

*p < 0.05. †p < 0.01. ‡p < 0.0001.

CHD = congenital heart defect; other abbreviations as in Tables 1 and 2.
Decreasing mortality associated with adult congenital heart disease


- Looked at trends in the US population from 1979 to 2005

In adults, overall decline in death rates in VSD, Ebsteins, coarctation and aortic stenosis (not specific for SCD)
Netherlands data

- 8595 ACHD patients, mean f/u 37 months
  - 2.7% mortality, mean age 48 years
  - 22% of deaths were sudden (<1% of population experienced SCD over this limited time frame)
    - 10% of these occurred during exercise
    - Ebsteins, DORV, TGA, VSD, aortic stenosis had more SCD than other lesions
      ("vascular" deaths not included in SCD)
Non-population based

• Toronto ACHD experience, 1981-1996
  – 2,609 patients
  – Data on 197/199 patients who died
    • 65% of deaths were cardiac
      – 26% sudden deaths (~2% of entire population)
    – New groups emerge in this older cohort:
      » Ebstein’s, L-TGA, as well as coarctation, AS and tetralogy

Oechslin, Am J Cardiol 2000;86:1111-16
Toronto series: mechanisms of sudden death in ACHD patients

- Non-cardiac: 33 (17%)
- Other cardiovascular: 36 (18%)
- Perioperative: 36 (18%)
- CHF: 41 (21%)
- SD: 51 (26%)
Lesion specific: tetralogy of Fallot

- Ghai et al, 2002: 12 ToF pts with SCD compared with 125 without SCD
- Pts with SCD more likely to have:
  - Moderate to severe PR
  - History of sustained ventricular tachycardia
  - QRS duration >180 msec
  - Left ventricular dysfunction

Combined positive predictive value 66%, negative predictive 93%

Multicenter tetralogy of Fallot data

- Gatzoulis et al: 793 tetralogy pts with no heart block or clinical arrhythmias
  - Mean time from repair 21 years, 10 yr window 1985-1995
  - 16 pts (2%) died suddenly
- Risk factors for SCD
  - Older age at repair
  - QRS duration >180 msec & increased rate of change in QRSd
  - Presence of tranannular patch

Lancet 2000 Sep 16;356(9234):975-81
Risk of sudden cardiac death in D-TGA with atrial switch surgery

**Overall SCD rates in adults ~5%**

  - 5.6% sudden deaths, no therapies in 5 ICD pts
- Roubertie et al 2011: 125 Senning survivors
  - 4% sudden deaths (4%)
  - 5% sudden late deaths, > late mortality in Mustard, VSD
468 Mustard/Senning patients, 1967-2003

- Follow-up in 2008
- After initial perioperative mortality, no significant differences in survival based on era of surgery, type of surgery, or associated defects
Conclusions

- Epidemiology data suggests higher risk of SCD in coarctation, aortic stenosis, tetralogy of Fallot and transposition, probably Ebsteins’, univentricular, L-TGA
- Death rates may be declining
- Some risk factors are apparent, best defined for tetralogy
- Constantly changing substrate
Thank you very much!