Sudden Cardiac Death and Malignant Arrhythmias
The Scope of the Problem in ACHD

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EP and Adult CHD Programs

Here’s a starting point...

Oechslin et al. *Am J Cardiol* 2000
2609 ACHD pts: 199 deaths (~8%) - sudden death (26%) and CHF (21%)
CoA (EF<40%), Ebstein (not WPW), ccTGA (EF<45%), AoV abnl, TOF

We haven’t understood this population very well...

Since late 1990’s, more CHD patients over age 18 than under in the U.S.

Now ~ 1,000,000 ACHD’ers
The SCD Problem in ACHD is a Continuum

Early events dictate and predict, to some extent, future risk of arrhythmia and SCD.

All of this leads to a substrate for late arrhythmia.

“30 years old” may be young, but it also means 25+ years exposure to post-op hemodynamics.
Categorizing the many different anatomies of ACHD:
Heart Failure leads to VT/SCD

“Dyssynchrony”
BBB, AVB, IVCD

Systemic RV Failure ↔ Systemic LV Failure ↔ Subpulmonary RV Failure

ccTGA, HLHS, Mustard/Senning, other Fontan
AI, CA abn’l, failed RV-LV interaction (TOF, Ebstein’s)
TOF, DORV

tachyarrhythmia

Sudden Cardiac Death

“Old-style Fontan” RA-PA connection

Current surgical management of “single ventricle physiology”

1 - Ao-Pulm shunt: imposes a volume load on the ventricle (“chronic” stretch)

2 - Bidirectional Glenn shunt: reduces volume, persistent low sats for myocardium

3 - Fontan: multiple suture lines in atria, atrial stretch
Many/most Fontan patients develop atrial tachyarrhythmias. Possibly decreasing incidence?

### Systemic SV Failure

Piran et al. *Circ* 2002;105:1189

long-term f/u of 188 adult CHD

**Adult Fontan patients:**

- 45% had sx’s of tachyarrhythmia or obvious CHF
- 18% died in 16 year follow-up
- 33% died 2° to HF
- ~90% of those dying with CHF had significant arrhythmia, mostly IART, VT

<table>
<thead>
<tr>
<th>Study</th>
<th>Method(s)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weber (AJC, 1989)</td>
<td>MIX at 10 yrs = 50%</td>
<td></td>
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<tr>
<td>Gelatt (JACC, 1994)</td>
<td>APC at 4½ yrs = 29%</td>
<td></td>
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<tr>
<td></td>
<td>TCPC = 14%</td>
<td></td>
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<tr>
<td></td>
<td>RV-PA = 18%</td>
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<tr>
<td>Cecchin (AJC, 1995)</td>
<td>APC at 7 yrs = 60%</td>
<td></td>
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<tr>
<td></td>
<td>LAT +/- FEN = 30%</td>
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<tr>
<td>Nürnberg (ATS, 2004)</td>
<td>early ECF/LAT = 11%, 38%</td>
<td></td>
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<tr>
<td>Bartz (JACC, 2006)</td>
<td>heterotaxy, 4 yrs = 50%</td>
<td></td>
</tr>
<tr>
<td>Giannico (JACC, 2006)</td>
<td>ECF at 15 yrs = 15%</td>
<td></td>
</tr>
<tr>
<td>Blaufox/PHN (JTCVS, 2008)</td>
<td>mixed = 9.6%</td>
<td></td>
</tr>
<tr>
<td>Stephenson/PHN (JACC, 2010)</td>
<td>all = 7.3%</td>
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</tbody>
</table>
**Tetralogy of Fallot**

Subpulmonary RV Failure

“tetralogy”
1. RV outflow obstruction
2. VSD
3. Aortic override
4. RVH

“monology”
Anterior deviation of the infundibular septum *in utero*

Infundibular incision

Pulmonary regurgitation
* contributes to RV failure (Frigola 2004)
* correlates with RV size, QRSd and VTs (Gatzoulis *Lancet* 2000, Abd El Rahman 2002)

QRS duration
* may be assoc. with VT (Gatzoulis 1997)
* improve after PVR (Therrien *Circ* 2002) - RVEDV 180ml/m²

Infundibular incision
* creation of isthmus-TA, VSD, PV (ablation data, Brigham, 2006)

Reentry ventricular tachycardia

SCD/VT in TOF - avg 27

avg 27 YEARS
VT in TOF

Counterclockwise VT circuit around the anastomosis of the RV and conduit.

Successful RF line created, posterior aspect of anastomosis and the tricuspid annulus.

Extreme bradycardia, sinus arrest on cessation of rapid atrial tachycardia. 25 yo with complex pulm atresia, failed 1 1/2 ventricle approach, atrio pulmonary Fontan
Bradycardia-induced VF converted by AED at a gym
**Systemic RV Failure** - Atrial Switch (Mustard, Senning)  
d-transposition of the great arteries

One operation. Good early result.

Long-term:  
Long atrial suture lines, conduction block  
Systemic right ventricle

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**Bradycardia**  
- “loss of sinus rhythm”  
- 20-60% at 20 yrs  
- pacing and HF  
- effects of slow HR on vent funet not well-studied  
- dilation due to increased stroke volume contributes to HF, VA's

**Atrial reentry**  
- most common tachy; can also 1:1 w/ ischemia, VT  
- rare paroxysm of adv AV block  
- uncontrolled IART: higher mortality risk

**1° VT/SCD**  
- Most ACHD w/ dTGA have fixed or reversible CA perfusion defects (systemic RV/coronary supply)  
- worse VT/SCD risk if Mustard with VSD  
- 70% have HF symptoms, poorly Rx’d  
- VT/VF documented in ~ all SCD, ~ 80% with exercise

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**Bradycardia/Tachycardias**  
d-TGA: Mustard, Senning

- 25 + \Rightarrow

<table>
<thead>
<tr>
<th>Bradycardia</th>
<th>Tachycardias</th>
</tr>
</thead>
<tbody>
<tr>
<td>NSR</td>
<td>AtrTachy</td>
</tr>
</tbody>
</table>

**Notes:**
- VT/SCD risk if Mustard with VSD
- 70% have HF symptoms, poorly Rx’d
- VT/VF documented in ~ all SCD, ~ 80% with exercise
**Coronary insuff:** Hornung. *Heart* 1998.

Pts with isolated cc-TGA
All w/ fixed perfusion defects, most w/ reversible defects
Only a 3 yr old had a normal EF

**CHF and SCD:** (age 45) Graham; Piran
- CHF in 67% with, 25% without assoc lesions
- >25% early sudden death, >1/2 with CHF
- VT/VF common w/ CHF - what’s a normal RVEF?

**Late AV Block**

“Natural” late AVB: ccTGA
- Risk of acquired AV block: 2%/yr, 7%/yr with VSD
  
  Huhta *et al.* *Circ* 1983

DILV, l-TGA (post-op Fontan)
- Risk of increasing degrees of AV conduction abnormalities over time

**Previous “transient” surgical AVB**
- 288 TOF pts, operated 1950-60’s
- 9% late sudden cardiac death
- Avg return of conduction in late SCD was POD 7.3
- Poor hemodynamics, later age at repair
- 7/7 discharged in SCAVB died
  
  Some with return of conduction 3-9 days can have late SCD (AVB)
  
  Hokansen, Moller. *Am J Cardiol* 2001
New AV block

- 57 yo Fontan, complex anatomy, AV-VA discordance
- 1st ECG in September
- 2nd ECG in October

Bradycardia and AVB-induced VT/VF
Ventricular tachycardia: LV failure

Systemic LV

Surgical intervention

avg 40 YEARS

RV-LV interaction:

AI and decr LV function after Ross operation:
- >15% require re-op for AI
- up to 30% may have early and/or late VT
- direct relationship between LVEDD and VA’s

RV-LV interaction:
- ****~10-15% of late post-op TOF patients will have reduced LVEF.
- Related to: 1) prior long duration shunt 2) aortic insufficiency 3) RV enlargement, reduced RVEF, RBBB
- Incidence of VA’s is higher in those with reduced LVEF
**Dyssynchrony**

**2**° to **Bundle Branch Block**

Adult HF: (CRT) Bradley et al. *JAMA* 2003;289;730
1634 pts, meta-analysis: CONTAK, MUSTIC, MIRACLE, InSync ICD

**LBBB 54-87%**, **LVEFs ≤ 30%**; Ischemic CM **37-69%**;
Age ~ 65 yrs

**ACHD**: RBBB - operations: TOF, VSD, etc, etc
LBBB - operations: AS, subAS, some DORV, AV canal

**2**° to **Pacing**

Adult pts: DAVID Trial (2002); MOde Selection Trial (2003)
Mortality, hosp and atrial fib > with more RV pacing

⇒ Over 1 year+

Ped/ACHD pts (mult studies)
incr. RV paced QRS duration and age = poorer LV function
Location: septum better than apex - LV dP/dt and LVEDP
“Upgrade” from to DDDR: Ventricular funct worse

⇒ Over decades?

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**The SCD Problem in ACHD is Personal**
The SCD Problem in ACHD is Changing
Changing… increasing complexity of the ACHD population

The problem of lifetime arrhythmia burden in ACHD needs to be quantified.
### CRM needs by CHD type

#### Estimate candidates:
- **Pacing:** AV block, SND/AT
- **ICD:** primary, secondary
- **CRT for HF**

<table>
<thead>
<tr>
<th>DEFECT</th>
<th>Incidence (in CHD Population)</th>
<th>Pediatric CRM needs</th>
<th>ACHD CRM needs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patent Ductus Arteriosus</td>
<td>7.3%</td>
<td>0.0%</td>
<td>0.0%</td>
</tr>
<tr>
<td>Pulmonary Stenosis</td>
<td>6.6%</td>
<td>0.0%</td>
<td>0.0%</td>
</tr>
<tr>
<td>Aortic Coarctation</td>
<td>6.2%</td>
<td>0.0%</td>
<td>6.0%</td>
</tr>
<tr>
<td>Ventricular Septal Defect</td>
<td>30.7%</td>
<td>2.5%</td>
<td>2.5%</td>
</tr>
<tr>
<td>Atrial Septal Defect</td>
<td>8.9%</td>
<td>6.0%</td>
<td>25.0%</td>
</tr>
<tr>
<td>Total Anomalous Pulmonary Venous Return</td>
<td>1.8%</td>
<td>2.0%</td>
<td>5.0%</td>
</tr>
<tr>
<td>Atrioventricular Canal</td>
<td>4.4%</td>
<td>5.0%</td>
<td>8.0%</td>
</tr>
<tr>
<td>Aortic Stenosis</td>
<td>4.0%</td>
<td>5.5%</td>
<td>38.0%</td>
</tr>
<tr>
<td>Truncus Arteriosus</td>
<td>1.9%</td>
<td>34.0%</td>
<td>26.0%</td>
</tr>
<tr>
<td>Pulmonary Abiesis</td>
<td>2.4%</td>
<td>3.0%</td>
<td>29.0%</td>
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<tr>
<td>Tetralogy of Fallot</td>
<td>4.9%</td>
<td>28.0%</td>
<td>42.0%</td>
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<tr>
<td>Double Outlet Right Ventricle</td>
<td>1.7%</td>
<td>14.0%</td>
<td>42.0%</td>
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<tr>
<td>Hypoplastic Left Heart</td>
<td>4.0%</td>
<td>14.5%</td>
<td>49.0%</td>
</tr>
<tr>
<td>Transposition of the Great Arteries</td>
<td>6.3%</td>
<td>34.0%</td>
<td>100.0%</td>
</tr>
<tr>
<td>OTHER - Single vent, ccTGA, heterotaxy, Ebstein’s</td>
<td>9.0%</td>
<td>32.0%</td>
<td>75.0%</td>
</tr>
</tbody>
</table>

**Pediatric CHD:** overall 9%

**Adult CHD:** overall 20%

< 10% are getting CRM Rx

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**The SCD Problem in ACHD needs a Plan**

[Image of a person]
Take home messages
Heart Failure and Arrhythmia in ACHD:
ACHD is an *electroanatomic* defect

- Important component of antiarrhythmic therapy in ACHD patients with HF is *heart failure therapy*.

- Anticipate EP needs by each CHD defect, physiology

- Use all available opportunities, don’t burn bridges

- Give the pediatric CHD patient a realistic appraisal of life as an adult CHD patient.