Arrhythmias in Adult Congenital Heart Disease

NAAMA’s 24th International Medical Convention
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June 26 – July 2, 2010

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I have no actual or potential conflict of interest in relation to this program or presentation. I will inform the audience of any off-label uses discussed.

Naser Ammash. MD
Arrhythmias in Adult CHD

Outline

1. Why do arrhythmias develop?
2. How common are arrhythmias?
3. Impact on patient’s outcome
4. Available treatment options
21 yr TGA S/P Arterial Switch
Near syncope at soccer field
Arrhythmias in Adult CHD
Ideal Substrate

Structural anatomic defect
Diseased myocardium
Residua + Sequelae
Large atria
Hypoxia
Scars

Inherent to CHD

Surgical sequelae
21 yr TGA S/P Arterial Switch
VT at soccer field

- Torsion, compression, intima thickening of CA
- Coronary obstruction
- Ischemia and ventricular arrhythmias
Tetralogy Of Fallot

30 yrs survival 86%
Occurrence of Malignant Arrhythmias in Tetralogy of Fallot

Patients 653

Survival 597

Deaths 56

Follow-up 23 years

Sudden 35%

CHF 33%

Other 34%

Murphy et al: NEJM 329:593, 1993
Saul: JCVEP 10:1271, 1999
Hemodynamic Substrate in for VT and SCD
456 Patients following TOF repair
Echo within 12 months

Analysis of QRS Duration vs RVEDV

37 post-TOF repair

\[ y = 0.18x + 99 \]

\[ r = 0.49, P < 0.01 \]

Plot of Maximum QRS Duration in Patients with Repaired Tetralogy of Fallot

Mechano - Electrical Interaction

Gatzoulis, Circulation, 1995
49 yr old S/P ASD surgical repair

Palpitation

...Valsalva
Secundum ASD
49 yr old S/P ASD surgical repair
Substrate

IART:
Macro reentry scar related
Slower than At. Flutter (<250 bpm)
Prone for 1:1 conduction
Adults with CHD have *many mechanisms* for atrial and ventricular arrhythmias that are *CHD specific*.
### Atrial Arrhythmias in Adult CHD

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Incidence of Arrhythmias</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>5-40% SVT</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>40-80% SVT</td>
</tr>
<tr>
<td>Single ventricle s/p Fontan</td>
<td>30-60% SVT</td>
</tr>
<tr>
<td>TGA s/p atrial switch repair</td>
<td>30-50% SVT</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>35% SVT</td>
</tr>
<tr>
<td>Tricuspid valve reoperation</td>
<td>35% SVT</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>&lt; 2% SVT</td>
</tr>
</tbody>
</table>
Lifetime Cumulative Incidence of Atrial Arrhythmias in ACHD

20-yr risk from age 20 = 7%

20-yr risk from age 55 = 38%

Adjusted cumulative incidence (% to develop AA if survived free of AA to age 18)

Age

Bouchardy et al: Circ 120:1679, 2009
Atrial Arrhythmia Adult CHD
Impact on Outcome

Any adverse event 2.50 (2.38-2.62)
Mortality 1.47 (1.37-1.58)
Morbidity 2.21 (2.07-2.36)
Stroke 1.55 (1.42-1.68)
Heart failure 2.64 (2.44-2.85)

HR for AA vs non-AA (95% CI)

Bouchardy: Circ, 2009
Proportion of Modes of CHD-Related Death in 6024 Post op survivors

<table>
<thead>
<tr>
<th>Mode</th>
<th>Number of Survivors</th>
<th>Incidence</th>
<th>Death</th>
<th>Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>PDA</td>
<td>2</td>
<td>13</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>COA</td>
<td>13</td>
<td>7</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>ASD</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>VSD</td>
<td>11</td>
<td>24</td>
<td>6</td>
<td>24</td>
</tr>
<tr>
<td>TOF</td>
<td>18</td>
<td>13</td>
<td>9</td>
<td>20</td>
</tr>
<tr>
<td>TGA</td>
<td>19</td>
<td>24</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>UVH</td>
<td>4</td>
<td>18</td>
<td>6</td>
<td>14</td>
</tr>
<tr>
<td>Misc</td>
<td>20</td>
<td>57</td>
<td>10</td>
<td>23</td>
</tr>
<tr>
<td>All</td>
<td>88</td>
<td>158</td>
<td>46</td>
<td>104</td>
</tr>
</tbody>
</table>

Incidence of arrhythmias *increases with age*

Significant *impact on outcome*

Drug Treatment of Adult CHD

- Proarrhythmia: Due to hypertrophy/dilatation
- Underlying sinus node dysfunction
- Conduction system defects
- One to one conduction in drug-organized atrial flutter
- Impairment ventricular function
- Altered drug metabolism: Renal/liver abnormalities
RFA of Atrial Arrhythmias in CHD

<table>
<thead>
<tr>
<th>Clinic</th>
<th>Successful Ablation</th>
<th>AT/AFLs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boston Childrens (n=10)</td>
<td>n=7 70%</td>
<td>22</td>
</tr>
<tr>
<td>Tanel/Boston Childrens (n=27)</td>
<td>n=16 59%</td>
<td>&gt;40</td>
</tr>
<tr>
<td>Kalman/UCSF (n=18)</td>
<td>n=15 83%</td>
<td>26</td>
</tr>
<tr>
<td>Van Hare/UCSF (n=10)</td>
<td>n=7 70%</td>
<td>13</td>
</tr>
</tbody>
</table>

Triedman: Circ 91:707, 1995
Van Hare: AJC 77:985, 1996
Numerous Mechanisms

Structural anatomic defect

Diseased myocardium

Residua + Sequelae

Large atria

Hypoxia

Scars

Success Rate: 85%
Tetralogy Of Fallot

Freedom from VT at 5 yrs in 70 rTOF: 100% with PVR + mapping / cryo

Therrien et al: Circ 103:2489, 2001
ACHD: Increased risk of SCD

- High Risk: TOF, dTGA, Fontan, LVOT obst., CoA, EA
- Technical challenges: Access, baffles, shunt, RVE
  - inferior epicardial lead performance due to scars
  - Coronary sinus, trans-myocardial apical
- Appropriate shock in 30% over 5 yrs FU
- Inappropriate shock in 25%, mostly due to ST
- Lead failure in 21%

Alexander et al, J Cardiovasc Electro 2004
Arrhythmias in Adult CHD

Conclusions

1. Many underlying mechanisms CHD specific
   • Residua or sequelae

2. Common: Atrial >> Ventricular
   • CHD specific
   • Incidence increase with age

3. Impact on patient’s outcome
   • CHF, stroke, hospitalization, QOL, SCD

4. Available treatment options
   • AA, DCCV, RFA, IOP Cryo or Maze, AICD
Age-adjusted death rate due to CHD

85% survive into adulthood

Projected Growth at 5% /yr

In the next decade, 1/150 adult will have CHD