Anomalous Aortic Origin of the Coronary Artery is Not Always a Surgical Disease

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AAOCA
Surgical Disease?

- Prevalence
- Risk of Sudden Cardiac Death / Complications
- Mechanisms (Anatomic Features) produce risk
  - Homogenous / heterogenous
- Reliability of Preoperative Testing
- Available Therapy
  - Reduce risk of SCD
  - Does intervention restore “normal” risk
  - Long term consequences of intervention
AAOCA - Prevalence

- Described over 2000 years ago by Galen
- Drawings “Tabulae Anatomicae” - Vesalius 1538
- True figures difficult to ascertain
- Multiple studies 0.1-0.3% (angiography)
- AAORCA 6x > AAOLCA
# Autopsy Study Data

<table>
<thead>
<tr>
<th>Author</th>
<th>N</th>
<th>AAORCA</th>
<th>AAOLCA</th>
<th>SCD- Right</th>
<th>SCD- Left</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chietlin-1974</td>
<td>51</td>
<td>18</td>
<td>33</td>
<td>0/18 (0%)</td>
<td>9/33 (27%)</td>
</tr>
<tr>
<td>Taylor-1992</td>
<td>30</td>
<td>21</td>
<td>9</td>
<td>4/21 (19%)</td>
<td>8/9 (89%)</td>
</tr>
<tr>
<td>Kragel-1988</td>
<td>32</td>
<td>25</td>
<td>7</td>
<td>8/25 (32%)</td>
<td>5/7 (71%)</td>
</tr>
<tr>
<td>Taylor-1997</td>
<td>101</td>
<td>52</td>
<td>49</td>
<td>13/52 (25%)</td>
<td>28/49 (57%)</td>
</tr>
<tr>
<td>Frescura-1998</td>
<td>11</td>
<td>7</td>
<td>4</td>
<td>4/7 (57%)</td>
<td>4/4 (100%)</td>
</tr>
<tr>
<td>TOTALS</td>
<td>225</td>
<td>123</td>
<td>102</td>
<td>29/123 (24%)</td>
<td>54/102 (53%)</td>
</tr>
</tbody>
</table>
Risk of Sudden Death

• Rates of SCD derived mainly from autopsy data
  – Mortality rates - 0-50% with AAORCA
  – 30-100% with AAOLCA

• Data are inherently biased by study population
  – Deceased
  – Reflection of prevalence of AAOCA in those who have died, **not** the risk of death of those living with anomalous coronary vessels.

Am Heart J 1997, 133:428-435
Hum Pathol 1998, 29:689-695
Similar Logic

- 35000 deaths in American males 15-24 yrs age
- 4200 deaths due to suicide
- Thus - risk of suicide is 12%
- Fortunately 1 in 8 males do not commit suicide
- US Census Data- (2014)
  - # males 15-24 years = 22,436,057
  - Actual risk = 0.0187%
SCD in US from AAOCA

- US Census (2014)
  - 318,892,103 total population
    - 0.1-0.2% of population affected
    - 315,000-630,000 people living with AAOCA
- Annual birth rate
  - ~ 4.5 million
  - 4000-8000 infants born each year with AAOCA
- Clearly if people were dying at rates predicted by autopsy series this would be an epidemic
## Rates of Sudden Death

<table>
<thead>
<tr>
<th>Author</th>
<th>Population studied</th>
<th>Total population (n)</th>
<th>Study period</th>
<th>Total sudden deaths</th>
<th>Cardiac related deaths</th>
<th>Deaths due to confirmed coronary anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wren et al., 2000</td>
<td>All children 1-20yo in Northern Health Region, England</td>
<td>806,000</td>
<td>1985-1994 (10 years)</td>
<td>270</td>
<td>26</td>
<td>(0%)</td>
</tr>
<tr>
<td>Eckart et al., 2004</td>
<td>All US military recruits</td>
<td>6,300,000</td>
<td>1977-2001 (25 years)</td>
<td>126</td>
<td>64</td>
<td>(.00003%)</td>
</tr>
<tr>
<td>Corrado et al., 2006</td>
<td>All people 12–35 years old in Veneto Region, Italy</td>
<td>4,379,900</td>
<td>1979-2004 (26 years)</td>
<td>N/A†</td>
<td>320</td>
<td>(.00004%)</td>
</tr>
<tr>
<td>Redelmeier and Greenwald, 2007</td>
<td>Marathoners from 26 selected US marathons</td>
<td>3,292,268</td>
<td>1975-2004 (30 years)</td>
<td>26</td>
<td>21</td>
<td>(.00006%)</td>
</tr>
<tr>
<td>Maron et al., 2009</td>
<td>All competitive US athletes</td>
<td>Not available</td>
<td>1980-2006 (27 years)</td>
<td>1866</td>
<td>1,049</td>
<td>NA</td>
</tr>
<tr>
<td>Chugh et al., 2009</td>
<td>All children 0-17yo in Multnomah County, OR</td>
<td>660,486*</td>
<td>2002-2005 (3 years)</td>
<td>8</td>
<td>3</td>
<td>(0%)</td>
</tr>
<tr>
<td>Harris et al., 2010</td>
<td>All triathletes in USA Triathlon sanctioned events</td>
<td>959,214</td>
<td>2006-2008 (3 years)</td>
<td>14</td>
<td>7†</td>
<td>(.00001%)</td>
</tr>
<tr>
<td>Harmon et al., 2011</td>
<td>All NCAA athletes</td>
<td>393,932§</td>
<td>2004-2008 (5 years)</td>
<td>80</td>
<td>45</td>
<td>N/A§</td>
</tr>
</tbody>
</table>

*Total population of Multnomah County, OR including children and adults.
†Officially listed cause of death “drowning” (during a triathlon event) but cardiac abnormalities were identified and thought to be the causative factor.
‡Study only looked at cardiovascular causes of sudden death.
§Total population derived by dividing “athlete participation years” by the 5-year study period. The study did not report on specific causes of cardiac death.

Penalver et al. BMC Cardiovasc Disorders 2012, 12:83
## Causes of Sudden Death in 387 Young Athletes

<table>
<thead>
<tr>
<th>Cause</th>
<th># of athletes</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic Cardiomyopathy</td>
<td>102</td>
<td>26.4</td>
</tr>
<tr>
<td>Commotio cordis</td>
<td>77</td>
<td>19.9</td>
</tr>
<tr>
<td>Coronary artery anomalies</td>
<td>53</td>
<td>13.7</td>
</tr>
<tr>
<td>LV hypertrophy of indeterminate causation</td>
<td>29</td>
<td>7.5</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>20</td>
<td>5.2</td>
</tr>
<tr>
<td>Ruptured aortic aneurysm (Marfan’s)</td>
<td>12</td>
<td>3.1</td>
</tr>
<tr>
<td>ARVD</td>
<td>11</td>
<td>2.8</td>
</tr>
<tr>
<td>Tunneled (bridged) coronary artery</td>
<td>11</td>
<td>2.8</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>10</td>
<td>2.6</td>
</tr>
<tr>
<td>Premature atherosclerosis</td>
<td>10</td>
<td>2.6</td>
</tr>
<tr>
<td>Dilated cardiomyopathy</td>
<td>9</td>
<td>2.3</td>
</tr>
<tr>
<td>Long QT syndrome</td>
<td>3</td>
<td>0.8</td>
</tr>
</tbody>
</table>

Maron BJ. JAMA 1996; 276:199-204
Facts- Summary

- Approximately 500,000 people USA are living with AAOCA
- AAORCA (1.3%) 6x > prevalence than AAOLCA (0.047%)
  - (Cath Cardiovas Diagn 1990;21:28-40)
- Sudden Cardiac Death (SCD) infants/ children
  - 0.8-6.2/100,000 per year
  - Coronary anomalies account for ~ 13%
    - (Circ 2009;119:1085-92)
- No patient SCD (AAOCA) was <10yrs or >30yrs age *
- Incidence of SCD with AAORCA rare
  - ~ 15 reported cases (1975-2015)
- Screening of active military recruits (n=6,3000,000)
  - 6,000-12,000 affected
  - 21 deaths due to AAOCA – all AAOLCA (risk = 0.17-0.35%)
    - (Ann Int Med 2004;141(829-34)

* 2 pts (1-9 yrs age) SCD while being treated medically- Ped Cardiol 2009, 30:911-921
Normal Anatomy
Normal?

Lack of consensus on definitions and diagnostic criteria

Based on anatomy or physiology

Clinical significance?

< 1% = abnormal

Angelini P- Circulation 2007;1296-1305
Possible Modes of Connection to “Opposite Sinus”

1. Retrocardiac
2. Retroaortic
3. Inter-arterial
4. Intraseptal / Intramural
5. Prepulmonary

Angelini P- Circulation 2007;1296-1305
AAOLCA Anatomy
Potential Mechanisms of Obstruction

1. Ostia- stenotic, slit-like
2. Acute angle of origin
3. Intramural course
4. Trans commissural course
5. Changes with exercise
   - Torsion
   - Compression
   - Vessel spasm
   - Intussusception

\[ F \propto \frac{\Delta P \cdot r^4}{\eta \cdot L} \]
Reliability of Preoperative Imaging

• Presence or Absence of Intramural Segment
  – Transthoracic Echocardiography (50-90%)
  – CT Angio (64-69%)
  – MRI (~83%)
Outcomes of Surgical Management of AAOCA

<table>
<thead>
<tr>
<th>Author</th>
<th>N (ARCA/ALCA)</th>
<th>Length of follow-up (mean or median in years)</th>
<th>Adverse outcome reported</th>
<th>Number of adverse events (ARCA/ ALCA)</th>
<th>Study limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Romp et al., 2003 [70]</td>
<td>9 (3/6)</td>
<td>2.4</td>
<td>Vavular dysfunction</td>
<td>1 (0/1)</td>
<td>Short follow-up</td>
</tr>
<tr>
<td>García-Rinaldi et al., 2004 [75]</td>
<td>16 (16/0)</td>
<td>6.8</td>
<td>Graft bleed, vessel occlusion</td>
<td>2 (2/0)</td>
<td>All adult patients (28-79yo) - No postoperative stress testing reported</td>
</tr>
<tr>
<td>Erez et al., 2006 [32]</td>
<td>9 (5/4)</td>
<td>1</td>
<td>None</td>
<td>0 (0/0)</td>
<td>Short follow-up</td>
</tr>
<tr>
<td>Alphonso et al., 2007 [74]</td>
<td>4 (2/2)</td>
<td>1.9</td>
<td>None</td>
<td>0 (0/0)</td>
<td>Short follow-up - Small N</td>
</tr>
<tr>
<td>Brothers et al., 2007 [56]</td>
<td>24 (16/8)</td>
<td>1.25</td>
<td>Ischemic changes, pericardial effusion</td>
<td>10 (8/2)</td>
<td>Short follow-up - No uniform preoperative testing</td>
</tr>
<tr>
<td>Fedoruk et al., 2007 [75]</td>
<td>5 (5/0)</td>
<td>1</td>
<td>RV dysfunction due to air embolus</td>
<td>1 (1/0)</td>
<td>Short follow-up - Small N</td>
</tr>
<tr>
<td>Gulati et al., 2007 [33]</td>
<td>18 (10/8)</td>
<td>2.2</td>
<td>Transient complete heart block, worsening heart failure, pericarditis requiring medical treatment</td>
<td>4 (Not specified)</td>
<td>Short follow-up - No postoperative stress testing reported</td>
</tr>
<tr>
<td>Hamzeh et al., 2008 [76]</td>
<td>4 (4/0)</td>
<td>2</td>
<td>Distal RCA stenosis requiring stent placement 48 hrs post-op</td>
<td>1 (0/1)</td>
<td>Short follow-up - Small N</td>
</tr>
<tr>
<td>Tavafi-Motamen et al., 2008 [61]</td>
<td>4 (4/0)</td>
<td>0.8</td>
<td>Graft failure</td>
<td>2 (2/0)</td>
<td>Short follow-up - Small N</td>
</tr>
<tr>
<td>Davies et al., 2009 [77]</td>
<td>36 (21/15)</td>
<td>1.1</td>
<td>Atrial fibrillation, subdural hematoma (died 2 months later)</td>
<td>7 (Not specified)</td>
<td>Short follow-up - No postoperative stress testing reported</td>
</tr>
<tr>
<td>el-Zein et al., 2009 [78]</td>
<td>8 (6/2)</td>
<td>1.2</td>
<td>None</td>
<td>0 (0/0)</td>
<td>- 2 patients lost to follow-up - Short follow-up - No postoperative stress testing reported</td>
</tr>
<tr>
<td>Fiomme et al., 2011 [79]</td>
<td>27 (20/7)</td>
<td>1.8</td>
<td>None</td>
<td>0 (0/0)</td>
<td>Short follow-up</td>
</tr>
<tr>
<td>Krasuski et al., 2011 [80]</td>
<td>28 (20/8)</td>
<td>8.2</td>
<td>None</td>
<td>0 (0/0)</td>
<td>- Outcome assessment limited to survival only</td>
</tr>
<tr>
<td>Mainwaring et al., 2011 [81]</td>
<td>48 (31/17)</td>
<td>5.7</td>
<td>Pleural effusions, postcardiomyopathy syndrome, heart block, heart transplantation</td>
<td>8 (Not specified)</td>
<td>- No postoperative stress testing reported</td>
</tr>
<tr>
<td>Munir et al., 2011 [82]</td>
<td>22 (15/7)</td>
<td>1.4</td>
<td>None</td>
<td>0 (0/0)</td>
<td>Short follow-up - Inconsistent postoperative stress testing</td>
</tr>
</tbody>
</table>

Adverse Events=36/238 (15%)
- chronic pericarditis
- pleural effusions
- postoperative bleeding
- heart block
- ischemic changes
- aortic valve insufficiency
- aortic valve replacement
- atrial fibrillation
- ostial stenosis
- graft failure
- CVA
- heart transplantation
- death

Abbreviations: ARCA = Anomalous right coronary artery from the left sinus; ALCA = Anomalous left coronary artery from the right sinus.
* Mortality was the only outcome assessed, all other studies listed reported on morbidity and mortality.
Reliability of Postoperative Testing

• Does Surgical Therapy?
  – Restore normal coronary flow
  – Allow unrestricted “Return to Play”
History - 15 year old male collapsed during basketball game
EKG - transmural anterolateral MI
ECHO - normal contractility, mild LVH
Troponin - 6.23ug/l (ref < 0.10ug/l)
Angio - inconclusive, ? AAOLCA
CMR - inconclusive, no delayed enhancement, LVEF-61%
CTA - AAOLCA with interarterial course

Surgery - AAOLCA, juxtacommissural, stenotic, slit-like orifice
LMCA ostial plasty and pulmonary artery translocation

Postoperative Course - medical therapy (metoprolol and aspirin)
Echo - good ventricular function, patent coronary artery
Stress Spect Test (52 mg adenosine, max 80 Watt load for 6 mins, MHR- 139bpm)
    no evidence of ischemia
Bicycle ergometry test - no evidence of arrhythmia or ischemia
SCD at basketball training that same afternoon - died despite aggressive resuscitation

Postop Mortality ~1.5%
Periop Testing - limited application, data and may be unreliable
Surgery may not allow for return to play status
Summary

- The prevalence of AAOCA is significant
- AAORCA >> AAOLCA
- The vast majority of patients go unrecognized and without negative consequences
- SCD very rare <10 yrs or >30 yrs age
- Multiple subgroups with variable and currently uncertain risk profiles.
- Surgical therapy appears to have low but finite risks
- Perioperative testing is inconclusive in determining risk stratification
Inferences- Recommendations

• Symptomatic AAOCA or evidence of ischemia: Surgical Intervention
• Asymptomatic AAOLCA: Surgical Intervention
• Asymptomatic AAORCA:
  – Therapy tailored toward assumed risk profile
    • Presumed activity level
    • Anatomic features of CA
      – Ostial characteristics
      – Intramural component
**Decision Algorithm**

**Symptoms**
- Ischemia

**Risk Profile**
- Ostium, Angle, Intramural, Cross Commissure

### AAOLCA
- **YES** → Surgery
- **NO**

### AAORCA
- **NO** → Surgery
- **Yes** → Concerning
  - **YES** → Surgery
  - **NO** → Observe
AAOLCA