Coronary Anomalies and Sudden Cardiac death in Athletes

Carlos M. Mery, MD, MPH
Texas Children’s Hospital
Sudden death of popular athlete stuns Pearland school

By Robert Stanton, Angel Verdejo | February 26, 2013 | Updated: February 28, 2013 10:47pm

Riverside High School Football Player Dies After Collapsing

The football player got up after a tackle, walked to the sideline and collapsed, a school spokesman said

By William Arla | Thursday Aug 29, 2013 | Updated 9:48 AM PST

Canyon High athlete, 14, dies after football practice

The teen collapsed during warm-ups for freshman football practice at the Anaheim school and died in the hospital Wednesday night
Should Young Athletes Be Screened for Heart Risk?

By ANAHAD O’CONNOR

Gary and Jocelyn Leonard during a tribute to their son, Wes, in Holland, Mich. Wes, 16, died on a basketball court moments after making a winning shot.
Outline

• Sudden cardiac death (SCD)
• Coronary anomalies / anomalous aortic origin of a coronary artery (AAOCA)
• Management of AAOCA patients
• Coronary Anomalies Program at TCH
Sudden Cardiac Arrest and Death

“Sudden cardiac arrest is the sudden cessation of cardiac activity so that the victim becomes unresponsive, with no normal breathing and no signs of circulation. If corrective measures are not taken rapidly, this condition progresses to sudden death.”

- ACC/AHA/HRS 2006 Guidelines
Age, race and gender related risk for SCD

Meyerburg et al. Cardiac Arrest and Sudden Cardiac Death in Braunwald’s Heart Disease: A Textbook of Cardiovascular Medicine, 2014:821-860.
Sudden Death in Young Athletes

Causes of Sudden Death in Young Athletes

Comparison of SCD Causes

ANOMALOUS ORIGINS OF THE CORONARY ARTERIES

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<table>
<thead>
<tr>
<th>KNOWNS</th>
<th>UNKNOWNS</th>
</tr>
</thead>
<tbody>
<tr>
<td>AAOCA is second cause of SCD</td>
<td>Mechanism of SCD?</td>
</tr>
<tr>
<td>AAOCA is common</td>
<td>Risk of SCD?</td>
</tr>
<tr>
<td>ARCA is more common but ALCA is more lethal</td>
<td>What characteristics are higher risk?</td>
</tr>
<tr>
<td>SCD usually during exercise</td>
<td>Optimal work-up?</td>
</tr>
<tr>
<td>50% have symptoms prior to SCD</td>
<td>Surgery?</td>
</tr>
<tr>
<td></td>
<td>Exercise restriction?</td>
</tr>
</tbody>
</table>
Counseling Patients

- Unclear disease pathogenesis
- Unclear treatment approach
- Unclear prognosis

Challenging counseling of patient & family
Treatment

Management of patients awaiting or not undergoing surgery

CHSS Survey (85 cardiologists, 24 CT surgeons, 4 others)

Brothers et al. Pediatr Cardiol 2009
Treatment

When to refer for surgical repair of AAOCA

CHSS Survey (85 cardiologists, 24 CT surgeons, 4 others)

Brothers et al. Pediatr Cardiol 2009
Asymptomatic ARCA: We are concerned that the risks of surgery outweigh the potential benefits and therefore advise against surgical intervention, but we recommend restriction from competitive sports or other recreational activities that approach the level of exertion seen in organized sports to further lower the risks. However, individual exceptions may be made if the risk of SCD associated with physical exertion is clear to the patient and that risk is deemed acceptable by both the patient and the cardiologist.
the important distinction between intramural and non-intramural course in asymptomatic patients with an anomalous right coronary currently cannot be made with definitive accuracy. It is also worth noting that while the prevalence of ischaemic symptoms is lower in non-intramural coronaries, half (4 of 8) of non-intramural right coronaries in our study were symptomatic. Because of this dilemma, we have recommended that all patients with AAOCA over the age of 10 years undergo surgical repair.
TCH Coronary Anomalies Program

• **Treat** patients with congenital coronary anomalies in an optimal and consistent way, based on the most current and reliable evidence available.

• **Educate** health care providers and the public about the latest information on risks, evaluation and treatment alternatives for patients with congenital coronary anomalies.

• **Study** the outcomes related to congenital coronary anomalies to improve the management of patients with this disease.

• **Engage** patients and families throughout the care management and longitudinal follow-up process to learn about their on-going experiences with this diagnosis.
Chest CT
Chest CT
Chest CT
Chest CT
Chest CT
Chest CT
Chest CT

Anomalous Coronary
Coronary Unroofing
Figure 1. CT scans of a patient undergoing coronary unroofing of an anomalous left coronary artery.

**Preoperative**

A) Anomalous left coronary artery (LCA, arrow) with an intra-arterial course and a 10mm intramural course.

B) Slit-like LCA ostium (*).

**Postoperative**

C) The left coronary has been unroofed, based on peri-coronary cuff of fat as well as the change in shape.

D) Improved caliber of the LCA ostium (*).
Coronary Translocation
Lateral PA Translocation

Anterior PA Translocation

Simplified Technique for Correction of Anomalous Origin of Left Coronary Artery From the Anterior Aortic Sinus

John M. Karamichalis, MD, Luca A. Vricella, MD, Daniel J. Murphy, MD, and Bruce A. Reitz, MD
Longitudinal Data Collection

Coronary Anomalies Program
Texas Children’s Heart Center

AAOCA Surgical Data Collection Form

Page 3-1

LEFT MAIN CORONARY ARTERY

☐ Origin from the aorta (fill below)
☐ Origin from another coronary (specify) _________

Location of ostium (select from each of the 3 sections - see Figures)

☐ 1: Right sinus of Valsalva
☐ X: Right/left commissure
☐ 2: Left sinus of Valsalva
☐ Y: Left/non-coronary commissure
☐ 3: ‘Non-coronary’ sinus of Valsalva
☐ Z: Right/non-coronary commissure

☐ a: Close to the first commissure
☐ b: Within ~50% of the center of sinus
☐ c: Close to the second commissure

☐ I: Centrally located
☐ II: High but below the sinotubular junction
☐ III: At the sinotubular junction
☐ IV: Above the sinotubular junction

Distance from commissure level to ostium: ________ mm
# TCH Coronary Anomalies Program

127 patients  
44 females (35%)  

<table>
<thead>
<tr>
<th>Presentation</th>
<th>ALCA</th>
<th>ARCA</th>
<th>Other</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Exertional symptoms</td>
<td>4 (17%)</td>
<td>25 (27%)</td>
<td>1 (8%)</td>
<td>30 (23%)</td>
</tr>
<tr>
<td>Symptoms at rest</td>
<td>7 (30%)</td>
<td>13 (14%)</td>
<td>3 (23%)</td>
<td>23 (18%)</td>
</tr>
<tr>
<td>Shock / SCD</td>
<td>4 (17%)</td>
<td>0</td>
<td>0</td>
<td>4 (3%)</td>
</tr>
<tr>
<td>Incidental finding</td>
<td>7 (30%)</td>
<td>46 (51%)</td>
<td>8 (62%)</td>
<td>60 (48%)</td>
</tr>
<tr>
<td>Total</td>
<td>23 (18%)</td>
<td>91 (72%)</td>
<td>13 (10%)</td>
<td>127</td>
</tr>
</tbody>
</table>

Data updated through 01/25/2017
Surgical Experience

43 patients (34%)
13 females (30%)
Median age: 14 years (8-18 years)

<table>
<thead>
<tr>
<th>Procedures</th>
<th>ALCA</th>
<th>ARCA</th>
<th>Total</th>
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</thead>
<tbody>
<tr>
<td>Unroofing</td>
<td>7</td>
<td>29</td>
<td>36</td>
</tr>
<tr>
<td>Coronary translocation</td>
<td>1</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Ostioplasty</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>9</td>
<td>34</td>
<td>43</td>
</tr>
</tbody>
</table>

Data updated through 01/25/2017
# Postop Symptoms and Clearance

<table>
<thead>
<tr>
<th></th>
<th>ALCA (n=9)</th>
<th>ARCA (n=34)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymptomatic</td>
<td>8 (89%)</td>
<td>32 (94%)</td>
</tr>
<tr>
<td>Non-specific CP</td>
<td>1 (11%)</td>
<td>2 (6%)</td>
</tr>
<tr>
<td>Cleared for exercise</td>
<td>8 (89%)</td>
<td>31 (91%)</td>
</tr>
<tr>
<td>Awaiting routine studies</td>
<td>1* (11%)</td>
<td>3 (9%)</td>
</tr>
</tbody>
</table>

Median follow-up: 1.8 years (1 month – 4 years)

* One after second intervention (LCA translocation and myocardial bridge)
SEPTEMBER 23 & 24, 2016
PHILADELPHIA, PA
Should Young Athletes Be Screened for Heart Risk?

By ANAHAD O’CONNOR

Gary and Jocelyn Leonard during a tribute to their son, Wes, in Holland, Mich. Wes, 16, died on a basketball court moments after making a winning shot.
Conclusions

- AAOCA is associated with higher risk of SCD
- The optimal management of AAOCA is controversial
- Standardized management by a dedicated multidisciplinary team provides consistent and optimal care for these patients
- Careful long-term follow-up of patients with AAOCA is necessary