ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERY

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ANOMALOUS

1. Inconsistent with or deviating from what is usual, normal or expected.

2. Of uncertain nature or classification.

3. Synonyms: irregular, unusual, equivocal or paradoxical

Webster’s Collegiate Dictionary
BOX 38-1  Congenital variations of the coronary artery

MINOR CORONARY VARIATIONS
High takeoff
Multiple ostia
Anomalous circumflex artery origin
Anomalous anterior descending artery origin
Absent proximal ostium/single ostium in other aortic sinus
Hypoplastic proximal coronary artery
Congenital proximal coronary artery
Congenital distal stenosis
Coronary artery from the posterior aortic sinus
Ventricular origin of an accessory coronary artery

MAJOR CORONARY ANOMALIES
Coronary “arteriovenous” fistula
Anomalous origin from the pulmonary artery
  Left coronary artery
  Right coronary artery
  Both coronary arteries

SECONDARY CORONARY ANOMALIES
Secondary coronary “arteriovenous” fistula
Variations in transposition of the great vessels
Variations in truncus arteriosus
Variations in tetralogy of Fallot
Distasia of coronary arteries in supravalvar aortic stenosis
Mural coronary artery

ALCAPA
AAOCA

is the acronym for
Anomalous Aortic Origin of a
Coronary Artery

by allacronyms.com
AORTIC ROOT ANATOMY

3 sinuses
Each named for respective coronary artery
<table>
<thead>
<tr>
<th>Type</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>LMCA from RASV</td>
<td>21%&lt;sup&gt;39&lt;/sup&gt; of AAOC: Four different courses possible in relation to</td>
</tr>
<tr>
<td>RCA from LASV</td>
<td>6% to 27%&lt;sup&gt;39,43&lt;/sup&gt; of AAOC: Can cross the aortic root anteriorly</td>
</tr>
<tr>
<td></td>
<td>pulmonary artery</td>
</tr>
<tr>
<td>CX from RASV or RCA</td>
<td>10% to 60%&lt;sup&gt;50&lt;/sup&gt; of AAOC (most common): Courses either ant vessels,</td>
</tr>
<tr>
<td></td>
<td>not between,&lt;sup&gt;50&lt;/sup&gt; always retroaortic&lt;sup&gt;51&lt;/sup&gt;</td>
</tr>
<tr>
<td>Inverted coronary arteries</td>
<td>Rare</td>
</tr>
</tbody>
</table>

CX, Circumflex coronary artery; LASV, left aortic sinus of Valsalva; LMCA, left main coronary artery; RASV coronary artery.
• Very contemporary
• Still being defined
• Controversies exist
• Different referral centers treat differently
  • Catheter based angiogram use to be the gold standard
  • CT angiogram may be now
• Some say Echo
AAOCA

Normal Origins

Abnormal Origin
RCA
AAOCA

• Left from right sinus

• Right from left sinus

• Rarely from non sinus
AAOCA FACTS

- Some types associated with sudden cardiac death (SCD)
- In the U.S., AAOCA is the second leading cause of SCD in children, after hypertrophic cardiomyopathy.
- SCD most commonly occurs during or just after exercise, notably among otherwise healthy young athletes

Maron et al, Circulation 1980;62: 218-219


Frescura C et al, Hum Pathol 1998; 29:689-95
AAOCA FACTS

Approx. 500,000 people USA living with AAOCA

AAORCA (1.3%) 6x> prevalence than AAOLCA (0.047%)


SCD children 0.8-6.2/100,000 per year

Coronary anomaly account for 13%

Circ 2009; 119: 1085-92

Ralph S Mosca, M.D.
No patient with SCD from AAOCA <10 yrs or >30 yrs*

*2 patients (1-9 yrs) SCD while being treated medically

Incidence of SCD with AAORCA rare approx 15 cases from 1975-2015

Ped Cardiol 2009;30: 911-921

Screening of active military recruits (n=6,300,000)

6,000-12,000 affected

21 deaths due to AAOCA-all AAOLCA (risk 0.17-0.35%)

Ann Int Med 2004; 141: 829-34

Ralph Mosca M.D.
## Autopsy Study Data

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

Intramural
SURGICAL TREATMENT

Interarterial
NOT SURGICAL VS SURGICAL

Anomalous LCA from right sinus - Inter-arterial Course
Increased risk of sudden death

Anomalous LCA from right sinus - Retro-aortic course
Prognosis benign
TRANSLOCATION
Ten-year experience with surgical unroofing of anomalous aortic origin of a coronary artery from the opposite sinus with an interarterial course

Peter C. Frommelt, MD, FACC, David C. Sheridan, MD, Stuart Berger, MD, Michele A. Frommelt, MD, FACC, and James S. Tweddel, MD

**Background:** Anomalous aortic origin of a coronary artery from the opposite sinus with an intramural course between the great arteries (AAOCA) is associated with ischemia and sudden cardiac death in children, and surgical unroofing has been used to alleviate that risk.

**Methods:** The cardiology database was reviewed to identify all patients with AAOCA who underwent surgical unroofing 10 years.

**Results:** From March 1999 to September 2009, 27 patients with a mean age of 12.6 ± 3.5 years (range, 4–22 years) had surgical unroofing of AAOCA of the left coronary from the right sinus (left AAOCA, 7/27, 26%) or of the right coronary from the left sinus (right AAOCA, 20/27, 74%). Of these, 26 had diagnoses made by transthoracic echocardiography. Symptoms included resuscitated sudden cardiac death in 3, syncope in 8, and chest pain in 4. No symptoms were present in 12 cases of serendipitous diagnosis. Unroofing of the intramural portion was successfully performed in all cases. A slitlike coronary orifice was described at surgical inspection in patients, 7 of whom had right AAOCA and no symptoms. All patients after unroofing have patent coronary flow by Doppler and normal echocardiography and exercise treadmill testing at mean follow-up of 1.8 years. None have activity restrictions.
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<td>8</td>
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<td>CT</td>
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<td>23</td>
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<td>TTE</td>
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</tr>
</tbody>
</table>

ADCA: Anomalous origin of coronary artery; Dx, diagnostic; CHD, congenital heart disease; F, female; LCA, left coronary artery; TTE, transthoracic echocardiogram; Cath, catheterization; CT, computed tomography; MRI, magnetic resonance imaging; TEE, transesophageal echocardiogram; AoV, aortic valve; VSD, ventricular septal defect.
• 27 patients with mean age 12.6 years

• Diagnosis of intramural course by echocardiography

• Treatment exclusively by unroofing

• 2/3 preop SCD on ecmo

• Left intramural course much longer then right

• All patients treated regardless of whether right, left asymptomatic, etc if intramural
NOW FOR THE MOVIE