The Second Leading Cause of Sudden Death in the Young Athlete: Coronary Anomalies

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I have no disclosures
“Though Sudden Cardiac Death (SCD) in the athlete is rare, its occurrence in athletes who are often young and presumably healthy has a large emotional and social impact on the surrounding community.”
The incidence of athlete deaths is not known with certainty, it would appear to be in the range of 1:200,000 high school age athletes per year.
Objectives

- Understand the anatomy of relevant coronary anomalies
- Review the prevalence of coronary anomalies
- Understand the pathophysiology of coronary anomalies
- Understand the diagnostic evaluation
- Review Treatment/interventions and controversy associated with this diagnosis
Normal Coronary Characteristics

<table>
<thead>
<tr>
<th>Feature</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of ostia</td>
<td>2 to 4</td>
</tr>
<tr>
<td>Location</td>
<td>Right and left anterior sinuses (upper</td>
</tr>
<tr>
<td></td>
<td>midsection)</td>
</tr>
<tr>
<td>Proximal orientation</td>
<td>45° to 90° off the aortic wall</td>
</tr>
<tr>
<td>Proximal common stem or trunk</td>
<td>Only left (LAD and LCX)</td>
</tr>
<tr>
<td>Proximal course</td>
<td>Direct, from ostium to destination</td>
</tr>
<tr>
<td>Mid-course</td>
<td>Extramural (subepicardial)</td>
</tr>
<tr>
<td>Branches</td>
<td>Adequate for the dependent</td>
</tr>
<tr>
<td></td>
<td>myocardium</td>
</tr>
<tr>
<td>Essential territories</td>
<td>RCA (RV free wall)</td>
</tr>
<tr>
<td></td>
<td>LAD (anteroseptal)</td>
</tr>
<tr>
<td></td>
<td>OM (LV free wall)</td>
</tr>
<tr>
<td>Termination</td>
<td>Capillary bed</td>
</tr>
</tbody>
</table>

LAD: Left anterior descending artery; LCX: Left circumflex artery; RCA: Right coronary artery; RV: Right ventricle; OM: Obtuse marginal artery; LV: Left ventricle.

**Dependent territory | Course | Branches**

<table>
<thead>
<tr>
<th>RCA</th>
<th>RV free wall</th>
<th>Right AV sulcus</th>
<th>At least the acute marginal branch</th>
</tr>
</thead>
<tbody>
<tr>
<td>LAD</td>
<td>Anterior IV septum</td>
<td>Anterior IV sulcus</td>
<td>Septal penetrating branches</td>
</tr>
<tr>
<td>LCX</td>
<td>LV free wall</td>
<td>Left AV sulcus</td>
<td>At least 1 obtuse marginal branch</td>
</tr>
</tbody>
</table>

RCA: Right coronary artery; LAD: Left anterior descending; LCX: Left circumflex artery; RV: Right ventricle; IV: Interventricular; LV: Left ventricle; AV: Atrioventricular.

*World J Radiol 2016 June 28; 8(6): 537-555 ISSN 1949-8470 (online)*
Table 1 Classification of anomalous coronary arteries

<table>
<thead>
<tr>
<th>1. Origin of both RCA and LMS (separate origins) from the right aortic sinus</th>
</tr>
</thead>
<tbody>
<tr>
<td>1a. Course of anomalous LMS between aorta and pulmonary artery (PA)</td>
</tr>
<tr>
<td>1b. Course of anomalous LMS not between aorta and PA</td>
</tr>
<tr>
<td>2. Origin of both coronary arteries (separate origins) from the left aortic sinus</td>
</tr>
<tr>
<td>2a. Course of anomalous RCA between aorta and PA</td>
</tr>
<tr>
<td>2b. Course of anomalous RCA not between aorta and PA</td>
</tr>
<tr>
<td>3. Anomalous origin of the circumflex coronary artery from the right aortic sinus</td>
</tr>
<tr>
<td>3a. Course of anomalous LCx between aorta and PA</td>
</tr>
<tr>
<td>3b. Course of anomalous LCx not between aorta and PA</td>
</tr>
<tr>
<td>4. Anomalous origin of the left anterior descending artery from the right aortic sinus</td>
</tr>
<tr>
<td>4a. Course of anomalous LAD between aorta and PA</td>
</tr>
<tr>
<td>4b. Course of anomalous LAD not between aorta and PA</td>
</tr>
<tr>
<td>5. Single coronary artery (common origin)</td>
</tr>
<tr>
<td>5a. Course of anomalous coronary artery between aorta and PA</td>
</tr>
<tr>
<td>5b. Course of anomalous coronary artery not between aorta and PA</td>
</tr>
<tr>
<td>6. Anomalous origin or communication of a coronary artery with a cardiac chamber or major thoracic vessel</td>
</tr>
<tr>
<td>6a. Abnormal origin from the pulmonary artery or one of its major arterial branches</td>
</tr>
<tr>
<td>6b. Abnormal origin from the aorta or one of its major arterial branches</td>
</tr>
<tr>
<td>6c. Abnormal communication of a coronary artery with a cardiac chamber or major thoracic vessel (fistula).</td>
</tr>
<tr>
<td>7. Miscellaneous/unclassified</td>
</tr>
</tbody>
</table>


Ripley et al. Journal of Cardiovascular Magnetic Resonance 2014, 16:34
http://jcmr-online.com/content/16/1/34
Anomalous Aortic Origin of a Coronary Artery (AAOCA)

- Typically refers to
  - Anomalous LCA from right sinus of Valsalva (ALCA)
  - Anomalous RCA from left sinus of Valsalva (ARCA)
  - Either from Non coronary sinus of Valsalva
  - Leading cause of sudden death from malformation, typically between the ages of 14-24
Anomalous Aortic Origin of the Coronary Artery (AAOC)

AAOCA: Course Subtypes
The 5 main course subtypes of anomalous aortic origin of a coronary artery (AAOCA) arising from the inappropriate sinus are shown: blue = pre-pulmonic; red = interarterial; orange = subpulmonic; green = retroaortic; purple = retrocardiac. Figure prepared by Robert Cheezum and Chris Shearin (DesignVis Studios Inc., Indianapolis, Indiana), and adapted with permission from Angelini et al. (80). Ao = aorta; MV = mitral valve; PV = pulmonic valve; TV = tricuspid valve.

Michael K. Cheezum et al. JACC 2017;69:1592-1608
Lethal Subtypes

- Anomalous left coronary from the right sinus of Valsalva
  - Inter-arterial course
  - Intramural course
- Anomalous right coronary artery from the left sinus of Valsalva
  - Inter-arterial course
  - Intramural course
- Anomalous Left coronary from the pulmonary artery
Anatomic Features

- **Ostia type:**
  - Separate
  - Shared
  - Branch

- **Take off angle:** 45-90 Degree angle vs. Sharp/Oblique slit like orifice

- **Intramural course (Within the wall of the aorta)**

- **Inter arterial course**
  - Oval shaped compressed vessel
Pathophysiology

- Myocardial infarction involving the territory of the anomalous coronary artery has been documented
  - Some studies suggest there have been multiple insults resulting in fibrosis prior to the fatal event
  - The infarcted heart muscle forms the substrate for malignant ventricular arrhythmias
Pathophysiology

- Slit like orifice:
  - During exercise:
    - Aortic engorgement
    - Systolic expansion of the aortic root
    - Valve like collapse of the orifice
    - Intimal damage and coronary spasm can result

_Congenital Anomalies of Coronary Arteries: Role in the Pathogenesis of Sudden Cardiac Death_

Melvin D. Chettilin, John MacGregor

Herz 2009;34:268-79
Pathophysiology

- Intramural course
  - The proximal portion of the vessel remains within the aortic wall at times for short distance other times for longer distances.
  - Aortic expansion in systole could compress the intramural portion

- Interarterial course
  - During vigorous activity engorgement of the great arteries could compress the anomalous vessel or cause kinking of the artery
<table>
<thead>
<tr>
<th></th>
<th>Echo</th>
<th>CTA</th>
<th>MRA</th>
<th>ICA</th>
<th>IVUS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Indication for AAOCA Imaging</strong></td>
<td>-</td>
<td></td>
<td></td>
<td>Class Ila</td>
<td>Class Ila</td>
</tr>
<tr>
<td><strong>Spatial Resolution</strong></td>
<td>0.8 × 1.5 mm (4-MHz transducer)</td>
<td>0.5 mm (isotropic)</td>
<td>1.0 mm (volumetric)</td>
<td>0.3 mm</td>
<td>0.15 × 0.25 mm</td>
</tr>
<tr>
<td><strong>Temporal Resolution</strong></td>
<td>30 msec</td>
<td>75-175 msec</td>
<td>60 - 120 msec</td>
<td>7-20 msec</td>
<td>Variable</td>
</tr>
<tr>
<td><strong>Visualize surround structures</strong></td>
<td>Limited</td>
<td>✔️</td>
<td>✔️</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td><strong>Dynamic imaging</strong></td>
<td>Limited</td>
<td>Limited</td>
<td>Limited</td>
<td>✔️ (Limited at ostium)</td>
<td>✔️</td>
</tr>
<tr>
<td><strong>Strengths</strong></td>
<td>✔ Noninvasive, rapid</td>
<td>✔ Noninvasive, rapid</td>
<td>✔ Noninvasive</td>
<td>✔ Noninvasive, rapid</td>
<td>✔ Dynamic imaging</td>
</tr>
<tr>
<td></td>
<td>✔ Widely available</td>
<td>✔ Visualize takeoff + course + surrounding structures</td>
<td>✔ Evaluate CAD</td>
<td>✔ Evaluate cardiac function, perfusion and prior MI</td>
<td>✔ Evaluation of proximal narrowing</td>
</tr>
<tr>
<td></td>
<td>✔ Low cost</td>
<td>✔ Evaluate multiple AAOCA features</td>
<td>✔ Avoid radiation &amp; iodinated contrast</td>
<td>✔ Invasive, Cost</td>
<td>✔ Invasive</td>
</tr>
<tr>
<td><strong>Limitations</strong></td>
<td>✔ Limited accuracy for detection of AAOCA</td>
<td>✔ Limited availability</td>
<td>✔ Limited availability</td>
<td>✔ Invasive, Cost</td>
<td>✔ Invasive</td>
</tr>
<tr>
<td></td>
<td>✔ Dependent on body habitus and operator technique</td>
<td>✔ Iodinated contrast</td>
<td>✔ Cost and scan-time increased vs. CTA</td>
<td>✔ Contrast and radiation</td>
<td>✔ Cost</td>
</tr>
<tr>
<td></td>
<td></td>
<td>✔ Radiation (low dose, e.g. 2-8 mSv now routine)</td>
<td>✔ Limited visualization of ostium, proximal course, surrounding structures</td>
<td>✔ Limited visualization of ostium, proximal course, surrounding structures</td>
<td>✔ Difficulty engaging anomalous vessel</td>
</tr>
</tbody>
</table>

Michael K. Cheezum et al. JACC 2017;69:1592-1608
1.5 year old with “seizures”

- Episodes of sudden loss of consciousness and stiffening, upward eye deviation and lipsmaking
- Also has breath holding spells
- Normal EEG
- Normal EKG
- Echo: Suspicious for AAORCA
- Dobutamine stress test: Max HR 191, BP 121/74: No evidence of ischemia
- Age 2.5 admitted for severe viral illness, again has “seizures”
  - Requires ST Lukes rehab
- Returns age 9: participates in long distance running, no chest pain but typically runs then walks then runs.
CPB 87 min
Cross clamp 69 min

7 mm intramural course unroofed

Discharged Post op day 2

Mild corneal abrasion
Post pericardotomy/pericardial effusion Treated with NSAID

Asymptomatic
Last seen 10/2017 Runs cross country
Due 10/2019
Presented age 12: Near syncope playing basketball
• During a scrimmage sudden onset of SOB while running, dizziness and extreme blurring of vision, difficulty hearing. She was “queasy”.
• Chest tightness and sharp pain with breathing
• ECG: IRBBB-Normal variant
• Stress ECG with nuclear med: Normal
Pre operative
- Surgical repair with unroofing procedure
  CPB 82 min
  Cross clamp 67 min
- 4 day hospital stay
- Non specific nuclear med changes, CT repeated and reassuring
- ASA 81 mg
- No exercise restriction
- 1/2017 (due for follow up)
  asymptomatic and normal Stress ECG
Intervention

- Exercise restriction
- Coronary bypass: not ideal. Competing flow results in bypass occlusion
- Coronary stent: odd position at the orifice
- Coronary unroofing/Osteoplasty: often requires resuspension of the aortic valve commisures
- Coronary transfer—rarely
Anomalous left coronary with long intramural segment

Anomalous left coronary with short intramural segment

Translocated Coronary
| TABLE 1. Clinical characteristics at presentation and intraoperative findings of patients with anomalous aortic origin of a coronary artery |
|--------------------------------------------------|-----------------|-----------------|-----------------|
|                                                   | Overall (n = 44) | ALCA (n = 9)    | ARCA (n = 35)   |
| Gender, female                                   | 13 (30)         | 3 (33)          | 10 (29)         |
| Age at surgery                                   | 14 y (8-18)     | 15 y (8-18)     | 13 y (8-18)     |
| Weight at surgery, kg                           | 53.7 (19.7-106) | 54.3 (23.92)    | 54.2 (19.7-106) |
| Asymptomatic                                     | 12 (27)         | 1 (11)          | 11 (31)         |
| Nonspecific symptoms                             |                 |                 |                 |
| Non-specific CP                                  | 16 (36)         | 4 (44)          | 12 (34)         |
| Syncope at rest                                  | 2 (5)           | 0               | 2 (6)           |
| Dizziness                                        | 9 (20)          | 1 (11)          | 8 (23)          |
| Palpitations                                     | 4 (9)           | 1 (11)          | 3 (9)           |
| Dyspnea on exertion                              | 3 (7)           | 1 (11)          | 2 (6)           |
| Exertional/ischemic symptoms                     |                 |                 |                 |
| CP on exertion                                   | 16 (36)         | 5 (55)          | 11 (31)         |
| Syncope on exertion                              | 7 (16)          | 4 (44)          | 3 (9)           |
| Aborted SCD                                      | 3 (7)           | 3 (33)          | 0               |
| Troponin leak/ECG changes                        | 1 (2)           | 0               | 1 (3)           |
| Hospital length of stay, d                       | 4 (3-10)        | 4 (3-6)         | 4 (3-10)        |
| CPB time, min                                    | 122 ± 35        | 119 ± 37        | 115 ± 34        |
| Crossclamp time, min                             | 82 ± 27         | 80 ± 23         | 77 ± 28         |
Surgical Outcomes

Frommelt et. al: University of Wisconsin:

10 year experience 1999-2009

27 pts mean age 12.6+-3.5 yrs (4-20) 7 (26%) AOLCA; 20 (64%) AORCA:

- Aborted SCD 3; syncope 8; chest pain 4

No deaths, short hospital stay for non SCD; all symptom free 1.8 years; 19 with normal post op treadmill and no exercise restriction
Who has AAOCA?

- Universal ECG screening?
- Universal Stress test screening?
- Universal echo screening?

Observed Prevalence of AAOCA

- Bias: evaluation typically occurs if there is a clinical indication for testing or serendipitously when getting tested for other reasons. Therefore do not really know the prevalence in the general population.

- Largest adult catheterization databank reported of 126,595 angiograms: ALCA 0.17%; ARCA 0.107%.

- Pediatric echo study: 14,546 echocardiograms: 24 (0.16%) ARCA; 6 (0.04%) ALCA (Congenital Anomalies of Coronary Arteries Role in the Pathogenesis of Sudden Cardiac Death Melvin D. Chetilin, John MacGregor1Herz 2009;34:268-79).

- Interartial: (Michael K. Cheezum et al. JACC 2017;69:1592-1608: Comprehensive review of published reports from 77 studies >1 million patients undergoing cardiac testing)
  - Anomalous LCA from Right sinus of valsalva 0.03% (3:10,000)
  - Anomalous RCA from Left sinus of valsalva 0.23% (2:1,000)
<table>
<thead>
<tr>
<th>First Author, Year (Ref. #)</th>
<th>Total Number of Autopsy Patients With ARCA/ALCA</th>
<th>Coronary-Related SCD</th>
<th>% of ARCA/ALCA Deaths During Exercise</th>
<th>% of ARCA/ALCA Asymptomatic Before Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cheitlin et al., 1974 (55)</td>
<td>ARCA 18, ALCA 33</td>
<td>ARCA 0 of 18 (0), ALCA 9 of 33 (27)</td>
<td>78</td>
<td>*</td>
</tr>
<tr>
<td>Kragel and Roberts, 1988 (56)</td>
<td>ARCA 25, ALCA 7</td>
<td>ARCA 8 of 25 (32), ALCA 5 of 7 (71)</td>
<td>*</td>
<td>38</td>
</tr>
<tr>
<td>Taylor et al., 1992 (57)</td>
<td>ARCA 24, ALCA 28</td>
<td>ARCA *</td>
<td>23 of 28 (82)</td>
<td>*</td>
</tr>
<tr>
<td>Taylor et al., 1997 (58)</td>
<td>ARCA 21, ALCA 9</td>
<td>ARCA 4 of 21 (19), ALCA 8 of 9 (89)</td>
<td>83</td>
<td>66</td>
</tr>
<tr>
<td>Frescura et al., 1998 (59)</td>
<td>ARCA 7, ALCA 4</td>
<td>ARCA 4 of 7 (57), ALCA 4 of 4 (100)</td>
<td>75</td>
<td>50</td>
</tr>
</tbody>
</table>

*Not reported. Adapted with permission from Mirchandani and Phoon (6).

ARCA = anomalous left coronary artery; ARCA = anomalous right coronary artery; SCD = sudden cardiac death.

<table>
<thead>
<tr>
<th>First Author, Year (Ref. #)</th>
<th>Population</th>
<th>N</th>
<th>Study Duration, yrs</th>
<th>Sudden Deaths</th>
<th>Deaths Attributed to CAA</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Total, n (%)</td>
<td>Cardiac, n</td>
</tr>
<tr>
<td>Wren et al., 2000 (81)</td>
<td>England, children 1 to 20 years of age</td>
<td>806,000</td>
<td>10</td>
<td>270 (0.03)</td>
<td>26</td>
</tr>
<tr>
<td>Eckart et al., 2004 (82)</td>
<td>U.S. military recruits</td>
<td>6,300,000</td>
<td>25</td>
<td>126 (0.002)</td>
<td>64</td>
</tr>
<tr>
<td>Corrado et al., 2006 (83)</td>
<td>Italy, population 12-35 yrs of age</td>
<td>4,379,900</td>
<td>26</td>
<td>*</td>
<td>320</td>
</tr>
<tr>
<td>Redelmeier and Greenwald, 2007 (84)</td>
<td>Runners from 26 U.S. marathons</td>
<td>3,292,268</td>
<td>30</td>
<td>26 (0.0008)</td>
<td>21</td>
</tr>
<tr>
<td>Maron et al., 2009 (85)</td>
<td>U.S. competitive athletes</td>
<td>*</td>
<td>27</td>
<td>1,866 (*)</td>
<td>1,049</td>
</tr>
<tr>
<td>Chugh et al., 2009 (86)</td>
<td>Oregon county, children ≤17 years of age</td>
<td>660,486†</td>
<td>3</td>
<td>8 (*)</td>
<td>3</td>
</tr>
<tr>
<td>Harris et al., 2010 (87)</td>
<td>U.S. triathletes</td>
<td>959,214</td>
<td>3</td>
<td>14 (0.001)</td>
<td>7</td>
</tr>
<tr>
<td>Harmon et al., 2011 (88)</td>
<td>NCAA athletes</td>
<td>393,932</td>
<td>5</td>
<td>80 (0.02)</td>
<td>45</td>
</tr>
</tbody>
</table>

*Not reported. †Total population of Multnomah County, Oregon, including children and adults. Adapted with permission from Peñalver et al. (89).

CAA = coronary artery anomalies (comprising all types of CAA; anomalous aortic origin of a coronary artery subtype not reported); NCAA = National Collegiate Athletic Association; SCD = sudden cardiac death.
Presenting symptoms

- Sudden Cardiac Arrest most frequently associated with or shortly after physical exercise
- The vast majority die in their teens or early 20s (age 14-24)
- Chest pain
  - Exertional
  - Post exertion
  - At rest
- Syncope/Pre-syncope
  - Exertional
  - Post exertion
- Family history
TABLE. The 12-Element AHA Recommendations for Preparticipation Cardiovascular Screening of Competitive Athletes

<table>
<thead>
<tr>
<th>Medical history*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Personal history</td>
</tr>
<tr>
<td>1. Exertional chest pain/discomfort</td>
</tr>
<tr>
<td>2. Unexplained syncope/near-syncope†</td>
</tr>
<tr>
<td>3. Excessive exertional and unexplained dyspnea/fatigue, associated with exercise</td>
</tr>
<tr>
<td>4. Prior recognition of a heart murmur</td>
</tr>
<tr>
<td>5. Elevated systemic blood pressure</td>
</tr>
<tr>
<td>Family history</td>
</tr>
<tr>
<td>6. Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in ≥1 relative</td>
</tr>
<tr>
<td>7. Disability from heart disease in a close relative &lt;50 years of age</td>
</tr>
<tr>
<td>8. Specific knowledge of certain cardiac conditions in family members: hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias</td>
</tr>
</tbody>
</table>

Physical examination
9. Heart murmur‡
10. Femoral pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position)§

*Parental verification is recommended for high school and middle school athletes.
Just because we can does that mean we should?

- We don’t truly understand the prevalence of the general population
- Is often diagnosed serendipitously in the adults
- Patient is most at risk during extreme exercise
- Patient is most at risk teenage to young adult years
- There is always a surgical risk
Recommended Guidelines

- Non-interarterial course
  - Conservative management, symptom directed

- Interarterial
  - No SX, no compression on CTA, neg Stress test
    - Conservative
    - Disqualify from competitive sports except ClassA1 (Bowling, cricket, golf)
  - Positive Sx, compression on CTA, + stress test
    - Intervention is justified:
      - Coronary unroofing
      - Coronary reimplantation
Future Directions

- CHSS registry
  - 1998-2009 Retrospective
  - 2009- Prospective

- ANOCOR
  - French multicenter study

- NIH
  - Sudden Death in the Young Registry
Secondary Prevention?

- At this time our best tool may be AED programs that make resuscitation possible
Figure 2 Kaplan-Meier curves showing the age at which the first major adverse cardiovascular event occurred, demonstrating a significant difference (log rank test, p < 0.0001) between those anomalous coronary arteries from the opposite sinus (ACAOS) with an inter-arterial course (IAC) and those without IAC.
In Summary

- SCD from coronary anomaly is rare
- Often young athletes are asymptomatic
- Not all with this diagnosis have the same risk for SCD
- Universal screening is not beneficial in identifying AAOCA
- 12 point Health questionnaire is our best tool at this time
  - Leads to many unnecessary echocardiograms
- Strategies for intervention are constantly changing
  - What may be state of the art now may create sequel 30 years from now
Coronary Anomalies: Anomalous Left Coronary artery from the Pulmonary Artery (ALCAPA)

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Embryology

- Early embryos have heart walls of loosely arranged muscle fibers/spongy subendocardium supplied with blood from the chambers.
- As the embryo gets older the heart walls thickened and muscle layers become more tightly packed. The intramyocardial sinusoids are precursors to intramural coronary arteries capillaries and veins.
- This particular anomaly may result from abnormal septation of the conal trunk vs persistence of pulmonary buds together with involution of aortic buds that eventually form the coronary arteries.
Pathophysiology

- Fetal life: Aorta and pulmonary artery oxygen saturations and pressures are similar therefore no harmful effect.
- Transitional circulation: Pulmonary artery contains desaturated blood and pulmonary artery pressures drop rapidly below systemic pressures.
- Left ventricle is perfused with desaturated blood at low pressure:
  - Left ventricular myocardial vessels dilate to reduce resistance and increased flow
  - Coronary vascular reserve becomes exhausted
  - Myocardial ischemia ensues
Figure 48.9 Left main coronary artery from pulmonary artery. (a) fetus, (b) in neonate. The light gray apical region indicates the ischemic region. (c) in older infant. The slightly darker gray apical region indicates worsening ischemia, and the very dark central area is the infarct. The gray vessels joining left anterior descending and right coronary arteries are collaterals. The pale gray arrows indicate the direction and approximate magnitude of collateral blood flow and its distribution. The open arrows indicate the direction and approximate quantity of flow in the proximal and distal left coronary artery.
Figure 30.11 Diagrammatic portrayal of varying functional states in anomalous origin of the left coronary artery from the pulmonary trunk. (a) During fetal life aortic (A) and pulmonary arterial (P) pressures are essentially equal. Flow in the anomalous artery is from the pulmonary trunk into the myocardium. (b) In early postnatal life pulmonary pressure has fallen below levels that pertain during fetal life. Rich intercoronary collateral channels have not yet developed. In this phase, flow through the anomalous coronary artery is probably at a low level. The anomalous vessel may be perfused either from the pulmonary trunk or from the right coronary artery through developing collateral systems. (c) In the final phase a rich collateral system has developed between the two coronary arteries. Characteristics of an arteriovenous fistula now pertain, with the major contribution to fistulous flow coming from the right coronary artery. Mediastinal arteries, which make communication with the coronary arterial system, may also contribute to such flow. Source: Adapted from Edwards JE. The direction of blood flow in coronary arteries arising from the pulmonary trunk. *Circulation* 1964;29:164, with permission of Wolters Kluwer Health.
Pathophysiology

Ischemia:
- At first ischemia is transient and occurs only with exertion
  - Crying or feeding
- Further increases in myocardial oxygen demand lead to infarction of the anterior lateral left ventricular free wall and Anterior papillary muscle of the mitral valve
- Heart failure ensues: Combination of myocardial infarction and mitral regurgitation
- Collateral vessels between the normal right and abnormal left coronary artery enlarge
- Coronary steal:
  - Right coronary artery → left coronary artery → pulmonary artery
Anterior lateral papillary muscle is atrophic and scarred with shortened/scarred chordae

Diffuse endocardial fibroelastosis of the left ventricle

Thinning and scarring of the anterior lateral ventricular wall and apex due to infarction, often mural thrombi as well

Cardiac enlargement especially involving the left ventricle and left atrium

Is usually isolated but has been associated with PDA, VSD, Tetralogy of Fallot, coarctation
Clinical features

- Completely asymptomatic until 6-10 weeks of age
- Paroxysmal episodes of acute discomfort/pain, diaphoresis, pallor with feeds.
- Progressing to tachypnea, decreased feeding, failure to thrive
- Some infants go on to have gradual improvement until they are asymptomatic
- Older children and adults may be asymptomatic or may have dyspnea and syncope and angina pectoris on exertion
- Sudden death after exertion is common
Clinical features

- Physical exam:
  - There may or may not be evidence of congestive heart failure at baseline.
  - The first heart sound may be soft or absent and there is mitral incompetence.
  - Loud second heart sound if left ventricular failure has caused pulmonary hypertension.
  - Apical gallop rhythm.
  - Murmur may be absent versus murmur of mitral incompetence.
  - A continuous AV fistula-type murmur may be present at the left upper sternal border.
  - Tachypnea, diaphoresis, hepatomegaly may be evidence of congestive heart failure.
Radiologic features

- Chest x-ray would demonstrate cardiomegaly primarily of left atrium and ventricle and evidence of pulmonary edema
Figure 48.10 Electrocardiogram in ALCA PA. Note abnormal q waves in leads V5 and V6, and also abnormal R wave progression in the precordial leads.
Echocardiographic features

- Cardiomegaly, especially left atrial and left ventricular enlargement
- Anterior lateral papillary muscle is usually ECHO Bright
- There may be ECHO Bright areas of the endocardium consistent with endomyocardial fibroelastosis
- Mitral regurgitation
- Left ventricular wall motion abnormalities
- Very large right coronary artery
- Color-flow demonstrating left coronary flow into the main pulmonary artery
Figure 13.1 (a) Echocardiogram in the parasternal short axis view showing normal origin of the left main coronary artery (LMCA) and its branches from the aorta. (b) Parasternal short axis view showing anomalous origin of the left main coronary artery (LMCA) from the pulmonary artery (PA), as seen in ALCAPA. Ao, aortic root; LAD, left anterior descending; LCX, left circumflex; LMCA, left main coronary artery; PA, pulmonary artery; RV, right ventricle.

Catheterization

- Echocardiography is now the diagnostic tool of choice
- Low cardiac output
- High filling pressures
- Angiography reveals dilated left ventricle and atrium and dysfunction of the anterior lateral left ventricular free wall and mitral regurgitation
- Aortic root angiography will demonstrated dilated right coronary artery, large collaterals and filling of the left coronary artery and passage of contrast into the pulmonary artery
Treatment

- The first effective surgical treatment was ligation of the origin of the left coronary artery to prevent steal; late sudden death could occur.
- Ligation of the origin of the left coronary artery with reconstitution of flow through a subclavian arterial or saphenous venous graft was successful but also at risk for clotting of the graft and Risk of late sudden death.
- Direct reimplantation of the origin of the left coronary artery to the aorta currently the procedure of choice.
- Alternative approach: Trapdoor flap method and tubular extension technique to minimize tension.
FIGURE 15-2
Intrapulmonary artery tunnel repair for anomalous origin of the left coronary artery (LCA) from the pulmonary artery (PA) (Takeuchi repair). A, Two dashed lines on the anterior wall of the PA are the proposed incision sites to create a flap of the PA. B, An aortopulmonary shunt is created after a punch hole (5–6 mm in size) is made in the contiguous wall of the aorta (Ao) and PA. C, The flap of the PA is sutured in place to form the convex roof of a tunnel through which aortic blood passes to the anomalous orifice of the left coronary artery. D, A piece of pericardium is used to close the opening in the anterior wall of the PA. E, Cross-sectional view of the tunnel operation when completed. LV, left ventricle; RA, right atrium; RV, right ventricle.
Treatment

- Mitral valvuloplasty often necessary
- Late survival for this surgery are fairly good approximating 90% at 5 years:
  - Heart becomes smaller
  - Heart failure abates
  - Mitral incompetence regresses
- Ongoing outpatient surveillance is important. At risk for development of diastolic dysfunction or ongoing mitral valve abnormalities
Clues to ALCAPA Diagnosis with echocardiography

- Congestive heart failure presentation in early infancy
- Severe left atrial and left ventricular dilation with abnormal systolic and diastolic function
- Left ventricular endocardial fibroelastosis changes
- Morphologically normal mitral valve with severe regurgitation
- Dilated Right coronary artery (RCA diameter to aortic root diameter ratio greater than 0.2)
- Retrograde flow into or 3 of coronary artery segments
- Prominent flow in septal perforators mimicking muscular VSD
- Abnormal diastolic flow jet in main or right pulmonary artery, increased with supplemental oxygen trial

Lai, W., Mertens, Luc, Cohen, Meryl, & Geva, Tal. (2016). Echocardiography in pediatric and congenital heart disease: From fetus to adult (Second ed.). Chichester, West Sussex; Hoboken, NJ: John Wiley & Sons.