WHAT THE REGISTRY TELLS US ABOUT AAOCA

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Marshall L. Jacobs MD
“AAOCA” with wrong sinus origin and interarterial +/- intramural course

Anomalous Aortic Origin of the LCA
Distribution of cardiovascular causes of sudden death in 1435 young competitive athletes

- HCM (36%)
- Indeterminate LVH - possible HCM (8%)
- Coronary artery anomalies (17%)
- Myocarditis (6%)
- ARVC (4%)
- MVP (4%)
- Tunneled LAD (3%)
- CAD (3%)
- AS (3%)
- Dilated C-M (2%)
- Sarcoidosis (1%)
- Aortic rupture (2%)
- Ion channelopathies (3%)
- Other congenital HD (2%)
- Other (3%)
- Normal heart (3%)

The true prevalence of AAOCA is unknown as there is no clear denominator. One large angiographic study evaluated 126,595 adults with coronary angiography and found the incidence to be approximately 0.3%\textsuperscript{1}

The largest population-based study in children using echocardiography found the incidence of AAOCA to be 0.17% in those with otherwise structurally normal hearts.\textsuperscript{2}


Recommendations:
1. Detection of coronary anomalies of wrong sinus origin in which a coronary artery passes between the great arteries should result in exclusion from all participation in competitive sports.
• AAOCA may be associated with SCD in young individuals, especially during or just after vigorous physical exertion
• Do all patients with “coronary anomalies of wrong sinus origin” need to refrain from competitive sports
• Are they “at risk,” even without competitive sports
• Which, if any of them, should have surgery?
• What are the ingredients and determinants of a successful operation?
Sudden Death as a Complication of Anomalous Left Coronary Origin From the Anterior Sinus of Valsalva

A Not-So-Minor Congenital Anomaly

By Melvin D. Chaitlin, M.D., COL, MC, Carlos M. De Castro, M.D., COL, MC, and Hugh A. McAllister, M.D., LTC, MC

SUMMARY

Both coronary arteries arising as a single or double vessel from the same sinus of Valsalva has been considered a minor congenital anomaly not affecting longevity. There are rare case reports in the literature of sudden death in young males with this anomaly of coronary origin. We have reviewed 51 such cases from the Armed Forces Institute of Pathology congenital heart disease accessions. There were 33 patients in whom both coronaries arose from the anterior sinus of Valsalva either as a single or double vessel and 18 in whom they arose from the left coronary sinus of Valsalva. Out of the 33 patients in whom the coronaries arose from the anterior sinus of Valsalva, 9 (27.3%) died sudden, unexplained deaths. There were no sudden unexplained deaths among the patients in whom both coronaries arose from the left sinus of Valsalva. It is evident that where the left coronary artery does not pass acutely posterior and leftward between the pulmonary artery and the aorta there is no risk of sudden death. All patients who died suddenly were male. The suggested mechanism for sudden death in these cases is that the acute leftward passage of the coronary artery along the aortic wall causes the entrance into the left coronary system to be slit-like. Under circumstances of increased cardiac activity with increased expansion of the pulmonary artery and aorta with exercise, there is stretching of the left coronary artery and a flap-like closure of the orifice of the left coronary with sudden, fatal myocardial ischemia.

We also present the first case where this anomaly was recognized in a 14-year-old boy and a surgical attempt was made to correct the problem by creating a non-collapsible funnel-like opening into the left coronary artery.
• 14 yo male presenting with ischemia and VT
  • Diagnosed with LMCA from anterior sinus

• Record of 17 yo male whose presenting event was sudden death after double-time march
  • LMCA from anterior sinus found at autopsy
  • Slit-like orifice and partial intramural course
Walter Reed Army Hospital

- Armed Forces Institute of Pathology: collected all autopsy cases with both coronaries arising as a single or double vessel from same sinus of Valsalva
  - 51 cases of AAOCA among 475,000 records
  - 33 patients with LMCA from anterior sinus
    - 9 of 33 (27.3%) with sudden, unexplained deaths
  - 18 patients with RCA from left coronary sinus
    - No cases of sudden cardiac death
Slit like orifice and intramural course was commonly noted and was the proposed mechanism of sudden death

Initial surgical plan for the 14 y.o. boy was coronary artery bypass

Ultimately performed operation: “Wedging out of the common wall and construction of a funnel-like opening to the left coronary system”

The first coronary unroofing operation: 1974
Sudden Death as a Complication of Anomalous Left Coronary Origin From the Anterior Sinus of Valsalva

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By Melvin D. Cheitlin, M.D., COL, MC, Carlos M. De Castro, M.D., COL, MC, and Henry A. McAnderson, M.D., LTC, MC

SUMMARY

Both coronary ostia have been considered a major cause of sudden death in the Armed Forces, in whom both coronary arteries arose from the anterior sinus of Valsalva... (i.e. ARCA from left sinus)
Issues

• Most cases described were sudden death or near deaths: How do we find it before sudden death?
• How rare? 33 out of the 475,000 AFIP autopsies
• No case under 12 years of age; this is congenital, are young children “at risk”?  
• Oldest 36 yrs; is there an age that it is no longer a problem?
• Is the mere presence of anomalous left coronary artery from the right (anterior) sinus of Valsalva an indication for surgery?
• Is the RCA from the left a benign condition?
Aberrant coronary artery origin from the aorta. Report of 18 patients, review of literature and delineation of natural history and management.

Nine patients had aberrant LCA. Three young males died suddenly after exertion, each with proximal focal LCA stenosis.

Nine patients (ages 18-60 years) had aberrant origin and course of the RCA. “Sudden death has not been reported with aberrant RCA, therefore in the absence of syncope, RCA bypass for sudden death prophylaxis is not indicated.”
Gaetano Thiene et al., Padua
Hum Pathol 1998;29:689-95.

Anomalous origin of coronary arteries and risk of sudden death: a study based on an autopsy population of congenital heart disease.

• 1200 congenital heart disease specimens studied
  • 11 cases of AAOCA (0.9%)
  • 4 of 4 cases of ALMCA w/ sudden death
  • 3 of 7 cases of ARCA w/ sudden death

• Conclusions:
  • ARCA can be lethal
  • AAOCA is not rare, though prevalence likely overestimated in this study
Autopsy study in young athletes


- 27 athletes who died suddenly during exercise
  - 24 ALMCA and 3 ARMCA
  - 22 men and 5 women, ages 9 to 32
  - 10 had symptoms (CP, syncope, palpitations)
  - 12 had normal testing in life (including all 10 symptomatic pt)
- Pathology: slit-like orifice, partially or completely intramural course in every case
Basso, Maron, et al.

Possible etiologies of sudden death in AAOCA

- Aortic pressure on intramural segment
- Slit-like orifice “closed” by increased aortic pressure
- Kinking of sharp angulation at take-off
- Inter-arterial segment “squeezed” by aorta and pulmonary artery
- Spasm of AAOCA
Risk of Sudden Cardiac Death reflected a complex interaction between the underlying morphology and the circulatory effects of intensive physical exertion.

Inference: geometry of coronary artery (including its ostium and proximal segment) may change *during strenuous exercise*.
EXERCISE-UNRELATED SUDDEN DEATH AS THE FIRST EVENT OF ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE RIGHT AORTIC SINUS

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*Pediatric Cardiology, Institute of Pediatrics, †Pediatric Intensive Care Unit, and ‡Department of Pathology, Catholic University of Sacred Heart, Rome, Italy
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- 10 year old girl no history of antecedent symptoms
- Died sitting at her desk at school
- ALMCA (single origin of both coronaries from the right sinus of Valsalva)
- Post-mortem evidence of past myocardial ischemia

Maybe Sudden Cardiac Death can occur unrelated to physical exertion

Acute ischemic event in the absence of exercise, OR Lethal arrhythmia initiated from previously injured myocardium
ARE ALL CASES OF AAOCA EQUAL?

ARE SOME MORE EQUAL THAN OTHERS?
RCA arising from the left coronary sinus and taking an inter-arterial course
Anomalous origin of RCA from left sinus of Valsalva with separate origin, interarterial course of RCA
Anomalous LCA from RCA, Interarterial

MRI View “from the foot of the bed”

Anomalous Origin of the Left Coronary Artery from the Right Sinus
Intraconal Coronary

From the head of the bed

From the foot of the bed
AAOCA includes a large number of “variations on a theme”

Unlikely that optimal treatment will be a “one size fits all” approach
AAOCA
Surgical repairs

- Unroof intramural segment (including creation of “neo-ostium”)
- Bypass (IMA or vein graft to affected coronary)
- Translocation of ostium (reimplantation)
- Translocation of pulmonary artery
- Revision (often pericardial patch plasty) of origin of anomalous coronary artery: alone or in combination with translocation of ostium or pulmonary artery
Surgical repair  AAOCA (RCA):
Unroofing intramural segment
Surgical repair AAOCA (LCA): Unroofing intramural segment
Surgical repair AAOCA: Unroofing intramural segment

13-year-old male with 2 episodes of angina & lightheadedness
LCA from right aortic sinus

C Mavroudis, R Stewart, CCF
Surgical repair AAOCA: Unroofing intramural segment

Probe demonstrates supra-commissural path
Surgical repair AAOCA: Unroofing intramural segment

Incision in common wall made with Potts scissors or no. 11 blade.
Surgical repair AAOCA: Unroofing intramural segment

Frequent use of probe to stay on path
Surgical repair AAOCA: Unroofing intramural segment

Edges of entire length of unroofed course is then tacked down with interrupted 8-0 Prolene.
Surgical repair AAOCA: Unroofing intramural segment

Final result, able to view coronary through wide open ostium.
Surgical repair AAOCA (LCA): Unroofing intramural segment
Surgical repair AAOCA (RCA):
Unroofing intramural segment with creation of neo-ostium
Surgical repair AAOCA (LCA):
Unroofing intramural segment with creation of neo-ostium
Modified Unroofing Procedure: Creation of neo-ostium in correct sinus
Modified Unroofing Procedure: Creation of neo-ostium in correct sinus
Simplified Technique for Correction of Anomalous Origin of Left Coronary Artery From the Anterior Aortic Sinus

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Department of Cardiothoracic Surgery and Division of Pediatric Cardiology, Stanford University School of Medicine, Stanford, California
Surgical Report: Duke
Turner et al., WJPCHS 2(3) 340-345; July 2011

- 53 patients
- 1995 - 2009
- Mean age 13.9 yrs (4-65 years)

- 40 ARCA, 13 ALCA

- Angina or syncope in 58% ARCA, 46% ALCA

- Lack of intramural course intra-op in 7 cases (5 ARCA, 2 ALCA)

- TTE accurately predicted intra/extramural in 92%

- No mortalities (mean F/U 29 months)

- One patient developed significant AR
## Table 2. AAOCA Morphology

<table>
<thead>
<tr>
<th>Morphology</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALCA Intramural</td>
<td>11 (84.6)</td>
</tr>
<tr>
<td>Extramural</td>
<td>2 (15.3)</td>
</tr>
<tr>
<td>ARCA Intramural</td>
<td>35 (87.5)</td>
</tr>
<tr>
<td>Extramural</td>
<td>5 (12.5)</td>
</tr>
</tbody>
</table>

Majority had intramural segment

Abbreviations: AAOCA, anomalous aortic origin of a coronary artery; ALCA, anomalous left coronary artery; ARCA, anomalous right coronary artery.

\(^a\) Percentage of total.
Table 4. Operative Procedures

<table>
<thead>
<tr>
<th>Operative Procedure</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Modified unroofing</td>
<td>43/53 (81.1)</td>
</tr>
<tr>
<td>Coronary translocation</td>
<td>6/53 (11.3)</td>
</tr>
<tr>
<td>Neoostial creation</td>
<td>3/53 (5.7)</td>
</tr>
<tr>
<td>CABG</td>
<td>1/53 (1.9)</td>
</tr>
</tbody>
</table>

Abbreviation: CABG, coronary artery bypass grafting.

a Percentage predictive of interarterial morphology based on intraoperative visualization and preoperative imaging modality obtained.
### Table 5. Surgical Outcomes

<table>
<thead>
<tr>
<th>Outcome</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Survival</td>
<td>No deaths</td>
</tr>
<tr>
<td>Complication rate (AI, PTX, pleural effusion, bleeding, retained foreign body)</td>
<td>5/53 (9.4)</td>
</tr>
<tr>
<td>Postoperative TTE patency</td>
<td>43/44 (97.7)</td>
</tr>
<tr>
<td>Postoperative symptomatic relief&lt;sup&gt;a&lt;/sup&gt;</td>
<td>35/43 (81.4)</td>
</tr>
<tr>
<td>Postoperative exercise restriction removal</td>
<td>36/40 (90)</td>
</tr>
</tbody>
</table>

Abbreviations: AI, aortic insufficiency; PT, pneumothorax; TTE, transthoracic echocardiography.

<sup>a</sup> There was no mortality in these 53 patients with 5 complications. Of the 8 patients who had some symptoms after surgery, only 4 had symptoms potentially consistent with cardiac ischemia and had negative functional testing for ischemia.
Surgical Report Stanford

Mainwaring RD, Reddy VM, Reinhartz O, Petrossian E, MacDonald M, Nasirov T, Miyake CY, Hanley FL.


• 50 patients
  • 1999 - 2010
  • Median age 14 yrs (5 days - 47 years)
  • 31 ARCA, 17 ALCA, 2 had eccentric single coronary ostium
  • 52% had symptoms of ischemia, 28% had associated CHD
  • Lack of intramural course intra-op in 7 cases (5 ARCA, 2 ALCA)
  • Repair by unroofing in 35, reimplantation in 6, PA translocation in 9
  • No operative mortality
  • Complications: PE 3, PCS 3, heart block 1
  • No SCD; median F/U 5.7 years
  • One Heart Transplant one year post-repair of AAOCA
Actuarial curves demonstrating survival (\{circ\}) and freedom from adverse event (\{square\}) for the 50 patients who underwent surgical repair of anomalous aortic origin of a coronary artery

Artist's illustration of the anterior pulmonary artery translocation procedure

Artist's illustration of the lateral pulmonary artery (PA) translocation procedure

Anomalous aortic origin of a coronary artery: a universally applicable surgical strategy

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Department of Cardiac Surgery, Queensland Paediatric Cardiac Service, Mater Children’s Hospital, Brisbane, Australia
Inferences

• SCD largely related to ALCA
• Ischemic symptoms and myocardial injury common in pts with ARCA
• Higher prevalence of associated CHD in ARCA group
• Pre-op symptoms correlated with intramural course, but not site of origin
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, a David C. Sheridan, MD, a Stuart Berger, MD, a Michele A. Frommelt, MD, FACC, a and James S. Tweddell, MD b

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, 4–20 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 12 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.

Conclusions: AAOCA is frequently characterized by an intramural course, which can be reliably identified by echocardiography. This form can be safely repaired by unroofing the intramural segment without early morbidity. Symptoms of possible ischemia are common but not always correlated with coronary ostial findings at surgery. (J Thorac Cardiovasc Surg 2011;142:1046-51)
Surgical Report, Milwaukee

- Patients:
  - 1999-2009
  - 27 patients
  - Mean 12.6 +/- 3.5 yrs
  - Range 4-20 yrs
  - AAOCA (left) 26%
  - AAOCA (right) 74%
  - All pts had 2 separate orifices within the same sinus
<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Sex</th>
<th>AOCA</th>
<th>Symptoms</th>
<th>Imaging Dx</th>
<th>Additional</th>
<th>Associated CHD</th>
<th>Ostial stenosis</th>
<th>Surgical technique</th>
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<tr>
<td>1</td>
<td>13</td>
<td>F</td>
<td>LCA</td>
<td>Syncope with exercise</td>
<td>TTE</td>
<td>Cath</td>
<td>None</td>
<td>Yes</td>
<td>Unroofing</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>M</td>
<td>LCA</td>
<td>None</td>
<td>TTE</td>
<td>Cath</td>
<td>Bicuspid AoV</td>
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<td>Unroofing; AoV resusp; VSD closure</td>
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<td>3</td>
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<td>Cath</td>
<td>VSD</td>
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<td>Unroofing; AoV resusp; VSD closure</td>
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<td>4</td>
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<td>M</td>
<td>LCA</td>
<td>SCD with exercise</td>
<td>TTE</td>
<td>TEE</td>
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<td>No</td>
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<tr>
<td>5</td>
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<td>M</td>
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<td>Cath</td>
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<td>No</td>
<td>Unroofing; AoV resusp</td>
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<td>Cath</td>
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<tr>
<td>7</td>
<td>10</td>
<td>M</td>
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<td>Nonexercise CP</td>
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<tr>
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<td>CP with exercise</td>
<td>TTE</td>
<td>Cath</td>
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<td>Yes</td>
<td>Unroofing</td>
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<td>None</td>
<td>None</td>
<td>Yes</td>
<td>Unroofing</td>
</tr>
<tr>
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<td>16</td>
<td>M</td>
<td>RCA</td>
<td>CP with exercise</td>
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<td>Yes</td>
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<tr>
<td>23</td>
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<td>CT</td>
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<td>None</td>
<td>No</td>
<td>Unroofing</td>
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<tr>
<td>27</td>
<td>11</td>
<td>F</td>
<td>RCA</td>
<td>Syncope with exercise</td>
<td>TTE</td>
<td>None</td>
<td>None</td>
<td>No</td>
<td>Unroofing</td>
</tr>
</tbody>
</table>

AOCA, Anomalous origin of coronary artery; Dx, diagnostic; CHD, congenital heart disease; F, female; LCA, left coronary artery; TTE, transthoracic echocardiogram; Cath, cardiac catheterization; M, male; AoV, aortic valve; RCA, right coronary artery; VSD, ventricular septal defect; resusp, resuspension; SCD, sudden cardiac death; TEE, transesophageal echocardiography; CP, chest pain; PVC, premature ventricular contraction; MRI, magnetic resonance imaging; CT, computed tomography; ECG, electrocardiogram; LVH, left ventricular hypertrophy; SVT, supraventricular tachycardia.
Intramural Coronary Length Correlates With Symptoms in Patients With Anomalous Aortic Origin of the Coronary Artery
*Ann Thorac Surg 2011;92:986-992*
DOI: 10.1016/i.athoracsur.2011.04.112

“A greater intramural coronary length measured by imaging or at surgery correlated with preoperative symptoms.”
Evaluation of Myocardial Ischemia Following Surgical Repair of Anomalous Aortic Origin of a Coronary Artery in a Series of Pediatric Patients

CONCLUSIONS— Subclinical changes suggestive of ischemia may occur despite patent neo-coronary ostia, notably after ARCA repair. The implication of these results on indication for surgery and subsequent sudden death risk is unknown. Serial EST, SE, and MPS are essential in evaluating ongoing ischemia risk after AAOCA repair.
Treatment Controversies

Treatment of AAOCA is controversial and varies among clinicians.

Recommendations may include observation alone, exercise restriction, medical therapy (e.g. beta blockers), and/or surgical repair.

Most agree that surgical intervention is indicated if a patient of any age presents with signs and/or symptoms of myocardial ischemia.

What remains unclear is the treatment of asymptomatic patients who are diagnosed serendipitously. The dilemma is greatest in those with ARCA, whose SCD risk is thought to be significantly lower than ALCA, and in young patients (i.e., ≤ 30 years) whose risk of SCD is probably greater than those identified in later adulthood.

Currently, we are unable to adequately risk stratify patients, to predict which are at higher risk for SCD, and to recommend evidence-based management choices.
Decision making should include consideration of specific features of morphology (imaging), clinical history, and physiologic testing for ischemia under conditions of stress.

Many centers advise surgery for most cases of ALCA and for ARCA only with symptoms c/w ischemia.
Unclear whether “negative” exercise study is reassuring, particularly in patients with ALCA.

Particularly unclear how, or if, asymptomatic patients with ARCA should be managed. (Gersony)
Management of Anomalous Coronary Artery From the Contralateral Coronary Sinus*

Welton M. Gersony, MD, FACC

New York, New York

At present, recommendations regarding exercise restriction as published in the American Heart Association/American College of Cardiology guidelines do not differentiate between ARCA and ALCA. These abnormalities are discussed together under the heading of Anomalous Coronary Arteries from the Contralateral Coronary Sinus (7). The profound difference in risk between the 2 sub-entities is not addressed. Thus, it is likely that, for a vast majority of asymptomatic patients with ARCA, excessive exercise limitations might be imposed, in many cases resulting in quality of life being unnecessarily affected.
Anomalous RCA: SCD In Athletes


29 y.o. **VF while playing basketball**, no prior hx. of symptoms
RCA from L sinus, separate orifice


39 y.o. elite runner, **VF during half-marathon**
RCA from L sinus, separate orifice, high, pericommisural, intramural
24 year old technical school instructor
VF while teaching, with no prodromic symptoms
CPR, defibrillation, rhythm restored but remained comatose
Ventilated, sedated and cooled for 48 hrs; awoke at 72 hrs
Cath: nl LMCA, **RCA from left sinus**
CT: **RCA from LMCA (common origin w/in left sinus)**, with intramural segment of RCA and “compression between Ao and PA”
Maybe we’re not so sure about the things we thought we were “pretty sure” about.

So, wouldn’t it be a good idea to gather more evidence?
"The unfortunate absence of national or international registers, which could provide precise data, suggests that SCD, although the present numbers are already impressive, is probably under-reported. The absence of precise data makes our/any analyses of reasons for SCD fragile and probably biased."
Anomalous Aortic Origin of a Coronary Artery with an Interarterial Course: Understanding Current Management Strategies in Children and Young Adults

Julie Brothers · J. William Gaynor · Stephen Paridon · Richard Lorber · Marshall Jacobs

“There is marked heterogeneity in physician opinions regarding AAOCA treatment and management.”

Cardiology in the Young (2010), 20(Suppl. 3), 50–58
doi:10.1017/S1047951110001095

Original Article

The registry of anomalous aortic origin of the coronary artery of The Congenital Heart Surgeons’ Society

Julie A. Brothers,1 J. William Gaynor,2 Jeffrey P. Jacobs,3 Christopher Caldarone,4 Anusha Jegatheeswaran,4 Marshall L. Jacobs,5 The Anomalous Coronary Artery Working Group
CHSS Registry of AAOCA

• Clinical Registry

• Imaging Registry

• “Virtual” Genomic Registry
The goal is to acquire and analyze a sufficiently large dataset that we will eventually be able to adequately risk stratify patients, to predict which ones are at higher risk for SCD, and to recommend evidence-based management choices.
CHSS Registry: Data Collection
<table>
<thead>
<tr>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>1. Sinus of Origin</td>
</tr>
<tr>
<td>2. # of Orifices</td>
</tr>
<tr>
<td>3. Dual Orifices</td>
</tr>
<tr>
<td>4. Slit-like orifice</td>
</tr>
<tr>
<td>5. High Take-off</td>
</tr>
<tr>
<td>6. Acute Angulation</td>
</tr>
<tr>
<td>7. Inter-arterial course</td>
</tr>
<tr>
<td>8. Intramural course</td>
</tr>
<tr>
<td>9. Intramural length</td>
</tr>
<tr>
<td>10. Intramyocardial</td>
</tr>
</tbody>
</table>
CHECKLIST: CHSS AAOCA

Date of Operation: ________________________

Which coronary is anomalous?  Right  Left
From which sinus does it arise?  Right  Left  Non-Coronal

Orifices:

- Two Separate orifices
- Separate Confluent orifices
- Single Orifice & bifurcation inside Aortic wall
- Single orifice & bifurcation outside Aortic wall

Is the orifice stenotic?  Yes  No
Diameter of the orifice: _____ mm
Is the orifice slit-like?  Yes  No
Location of AAOCA orifice:  Normal
- High & at commissure
- High & below commissure
- High & above commissure

Please indicate the location of the orifice by drawing it on this diagram
Schematic representation of the possible origin of coronary arteries' orifices according to the vertical and radial orientation relative to the sinuses and to the comissures of the aortic valve.

Aiello V D,  World Journal for Pediatric and Congenital Heart Surgery 2010;2:9-18
DIAGRAM 3
Example of an angulated orifice: The coronary vessel passes obliquely through the aortic wall for a distance which is less than the diameter of the coronary artery.

Length of intramural course

Ostium

Aorta

DIAGRAM 4
Example of an intramural course: The coronary vessel passes obliquely through the aortic wall for a distance which is greater than the diameter of the coronary artery.

Length of intramural course

Ostium

Aorta
CT Scan

“navigator view”
Reconstructed three-dimensional computed tomographic (CT) scan viewed from the surgeon's perspective looking down toward the aortic valve

Baird  C. W.; Ann Thorac Surg 2010;89:1341
The CHSS AAOCA Registry

Patients $\leq$ 30 years of age
Diagnosis of AAOCA at a CHSS institution after 1998

Excludes patients with other clinically important cardiac lesions

N=197

Thanks to Jeffrey Poynter MD, CHSS Kirklin-Ashburn Fellow
Participating Centers
Interim Results
Flow Diagram

AAOCA
N=197

Right
N=143

Both
N=3

Left
N=51

Based on imaging studies
Symptoms

**Symptomatic**

- Chest pain during exercise: 45%
- Chest pain at rest: 28%
- Syncope: 17%
- Other: 10%

**Asymptomatic**

- Screening for murmur
- Screening for family history
Flow Diagram

AAOCA
N=197

Right
N=143

Both
N=3

Left
N=51

Observation

Surgery
Flow Diagram

AAOCA
N=197

Right
N=143

Both
N=3

Left
N=51

Observation
2
20
1
31

Surgery
Flow Diagram

- AAOCA
  - N=197
    - Right
      - N=143
    - Both
      - N=3
    - Left
      - N=51
- Observation
  - N=93
- Surgery
  - N=104
### Flow Diagram

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<th>Category</th>
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<td><strong>Surgery</strong></td>
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<tr>
<td>Inter-arterial and Intra-mural</td>
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<tr>
<td>Inter-arterial Only Only</td>
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<tr>
<td>Other</td>
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<td>Inter-arterial Only Only</td>
<td>58</td>
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<td>Other</td>
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## Primary Operations

<table>
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<tr>
<th>Procedure</th>
<th>Inter-arterial and intra-mural (N=95)</th>
<th>Inter-arterial without intra-mural (N=8)</th>
<th>Not inter-arterial or intra-mural (N=1)</th>
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<tbody>
<tr>
<td>Unroofing</td>
<td>82</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ostial Window</td>
<td>5</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Reimplantation</td>
<td>8</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Pulmonary Artery Translocation</td>
<td>0</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>CABG</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Non-operative Controls Available</td>
<td>26</td>
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<td>6</td>
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Correlations

<table>
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<tr>
<th>Symptomatic</th>
<th>% Surgery</th>
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<tr>
<td>Yes</td>
<td>65%</td>
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<tr>
<td>No</td>
<td>44%</td>
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</table>

<table>
<thead>
<tr>
<th>Inter-arterial and Intra-mural Course</th>
<th>% Surgery</th>
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</thead>
<tbody>
<tr>
<td>Yes</td>
<td>66%</td>
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<tr>
<td>No</td>
<td>45%</td>
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<table>
<thead>
<tr>
<th>Inter-arterial Course Only</th>
<th>% Surgery</th>
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<tbody>
<tr>
<td>Yes</td>
<td>47%</td>
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</table>
Conclusions

Overall, 51% of patients with ARCA and 71% with ALCA had surgery.

Most patients referred for surgery had an intramural course of the anomalous coronary artery
Inferences

There is significant variability in management practices

Surgical intervention is weakly correlated with symptoms and morphology

Long-term outcomes will allow us to establish evidence-based guidelines
Thank you for staying with me...
Q17. Anomalous origin of the left coronary artery is associated with sudden cardiac death, but anomalous origin of the right coronary is not. True or False?

a. True
b. False
Q18. The most common surgical procedure performed for AAOCA in children is bypass grafting with internal thoracic artery. True or False?

a. True
b. False
Q19. Anomalous origin or the left coronary artery with intraconal course is generally considered to be associated with a high risk of sudden cardiac death.

True or False?

a. True
b. False
<table>
<thead>
<tr>
<th>Anomalous Coronary Artery</th>
<th>No. Interarterial (%)</th>
<th>No. Intramural (%)</th>
<th>No. Intraconal (%)</th>
<th>No. At or Above Commissure (%)</th>
<th>No. Below Commissure (%)</th>
<th>No. Slit-like Orifice (%)</th>
<th>No. Stenotic Orifice (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left</td>
<td>32 (28%)</td>
<td>24 (75%)</td>
<td>29 (91%)</td>
<td>1 (3%)</td>
<td>27 (84%)</td>
<td>2 (6%)</td>
<td>11 (34%)</td>
</tr>
<tr>
<td>LAD</td>
<td>1 (1%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>1 (100%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Right</td>
<td>79 (69%)</td>
<td>70 (89%)</td>
<td>75 (95%)</td>
<td>0 (0%)</td>
<td>68 (86%)</td>
<td>8 (10%)</td>
<td>42 (53%)</td>
</tr>
<tr>
<td>Both</td>
<td>2 (2%)</td>
<td>2 (100%)</td>
<td>1 (50%)</td>
<td>0 (0%)</td>
<td>2 (100%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
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</table>

**Operative Detail**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>N</th>
<th>%</th>
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</thead>
<tbody>
<tr>
<td>Unroofing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>with excision of tunica media and intimal tacking</td>
<td>99</td>
<td>87</td>
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<tr>
<td>with excision of tunica media without intimal tacking</td>
<td>26</td>
<td>26</td>
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<tr>
<td>with incision of tunica media and intimal tacking</td>
<td>47</td>
<td>47</td>
</tr>
<tr>
<td>with incision of tunica media without intimal tacking</td>
<td>13</td>
<td>13</td>
</tr>
<tr>
<td>PA translocation</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>CABG</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>with internal mammary artery graft</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>with proximal coronary artery ligation</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Aorto-coronary window</td>
<td>6</td>
<td>5</td>
</tr>
<tr>
<td>Ostioplasty</td>
<td>29</td>
<td>25</td>
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<tr>
<td>with patch augmentation</td>
<td>14</td>
<td>12</td>
</tr>
<tr>
<td>Commissure takedown</td>
<td>37</td>
<td>32</td>
</tr>
<tr>
<td>with subsequent resuspension</td>
<td>33</td>
<td>29</td>
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</table>
There are three basic categories. First, minor anomalies in which there is a variation of the origin of the vessels from the aorta and the distal circulation is normal………
<table>
<thead>
<tr>
<th>Anomalous Coronary Artery</th>
<th>N (%)</th>
<th>No. Interarterial (%)</th>
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<td>68 (86%)</td>
<td>8 (10%)</td>
<td>42 (53%)</td>
<td>12 (15%)</td>
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<td>Both</td>
<td>2 (2%)</td>
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<td>2 (100%)</td>
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<tr>
<td>Operative Detail</td>
<td>N</td>
<td>%</td>
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<td>Unroofing</td>
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<td>Ostioplasty</td>
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<td>with patch augmentation</td>
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# Primary Operations

<table>
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<tr>
<th>Procedure</th>
<th>Inter-arterial and intra-mural (N=95)</th>
<th>Inter-arterial without intra-mural (N=8)</th>
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<tr>
<td>Unroofing</td>
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<td>Ostial Window</td>
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<td>Reimplantation</td>
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<td>CABG</td>
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<tr>
<td>PA translocation</td>
<td>8</td>
<td>8</td>
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</tbody>
</table>
Frommelt, Milwaukee
Observations and inferences

- 12 of 27 were without symptoms of ischemia (found on TEE for suspicion of CHD – present in 4 of 12)
- L AAOCA: all had intramural course, 1 had slit-like orifice
- R AAOCA: shorter intramural course, orifice near commisure, 11/20 slit-like orifice
- Etiology of ischemia multi-factorial; not always explained by slit-like ostium or angulation
- *Symptoms do not always correlate w/ ostial findings*
A Few Words of Caution

Coronary Steal Syndrome After Coronary Artery Bypass for Anomalous Aortic Origin of a Coronary Artery

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Anomalous aortic origin of a coronary artery found in a symptomatic 9-year-old boy was initially treated with coronary artery bypass grafting using a left internal mammary artery anastomoses to the left anterior descending coronary artery, but resulted in coronary ischemia, likely from a steal phenomenon. Subsequent transection of the proximal left internal mammary artery with anastomosis to the ascending aorta, and coronary ostial enlargement, resulted in a durable treatment. We recommend caution in choosing coronary artery bypass grafting using a left internal mammary artery pedicle graft for the treatment of anomalous aortic origin of a coronary artery.

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Fig 1. Initial postoperative coronary angiogram showing anomalous origin of the left coronary artery from the right coronary sinus of Valsalva. Retrograde flow from the left anterior descending coronary artery to the left internal mammary artery to the left subclavian artery was seen (arrow shows direction of flow).