Symptoms and Coronary Anatomy are Associated with Management Decisions in Children and Young Adults with Anomalous Aortic Origin of a Coronary Artery: A Congenital Heart Surgeons’ Society Study

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Background

Anomalous aortic origin of a coronary artery (AAOCA) is the 2nd leading cause of sudden cardiac death in children & young adults

No evidence-based management guidelines

Morphology varies widely
AAOCA Variant:
Inter-Arterial Anomalous LCA
AAOCA Variant: Inter-Arterial and Intra-Mural
Slit-like Orifice

ARCA

LCA

Slit-like ARCA Orifice

Commissure

Round LCA Orifice

Courtesy of Matt Harris, MD
Long-term Objectives

Develop a registry

Describe the natural and “unnatural” history of AAOCA

Determine risk factors

Create evidence-based guidelines for management
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
Participating Centers
Interim Results
Flow Diagram

AAOCA
N=197

Right
N=143

Both
N=3

Left
N=51
Symptoms

Symptomatic  54%
  Chest pain during exercise  45%
  Chest pain at rest  28%
  Syncope  17%
  Other  10%

Asymptomatic  40%
  Screening for murmur  56%
  Screening for family history  44%
Flow Diagram

AAOCA
N=197

Right
N=143

Both
N=3

Left
N=51

71  Observation

72  Surgery
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
N=93

Surgery
N=104
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<thead>
<tr>
<th>Category</th>
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<th>Surgery</th>
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<tbody>
<tr>
<td>Inter-arterial and Intramural</td>
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<td>N=50</td>
</tr>
<tr>
<td>Only</td>
<td>N=51</td>
<td>N=58</td>
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<tr>
<td>Other</td>
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**Flow Diagram**

N=104

N=93
## Correlations

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<tr>
<th>Symptomatic</th>
<th>% Surgery</th>
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<td>Yes</td>
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<table>
<thead>
<tr>
<th>Inter-arterial and Intra-mural Course</th>
<th>% Surgery</th>
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<tbody>
<tr>
<td>Yes</td>
<td>66%</td>
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<tr>
<td>No</td>
<td>45%</td>
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<tr>
<th>Inter-arterial Course Only</th>
<th>% Surgery</th>
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<td>Yes</td>
<td>47%</td>
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<tr>
<td>No</td>
<td>60%</td>
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Conclusions

The CHSS has established a comprehensive registry of young patients with AAOCA.

Surgical intervention is weakly correlated with symptoms and morphology.
Inferences

There is significant variability in management practices

Long-term outcomes will allow us to establish evidence-based guidelines
Enrollment
We continue to enroll AAOCA patients from CHSS institutions

Your help with requests for follow-up information will be appreciated
Acknowledgment

Our study is funded by generous contributions from:
The Children’s Heart Foundation,
The Cardiac Center at The Children’s Hospital of Philadelphia and
Congenital Heart Surgeons’ Society (CHSS) member institutions
Present Study Objectives

Outline inclusion criteria, recruitment, demographics, clinical findings and triage

Report symptoms, coronary morphology and interventions

Examine agreement between diagnostic image reports and surgical reports
Agreement: Imaging vs. Surgery

Good or excellent agreement ($\kappa > 0.7$, $p < 0.001$) for:

- Which CA is anomalous
- Slit-like orifice
- Acute angulation
- Intramural segment
- Interarterial course
- Sinus (or site) of origin
### Flow Diagram - Surgical Views

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<th>Surgery</th>
<th>Observation</th>
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<tbody>
<tr>
<td></td>
<td>N=104</td>
<td>N=93</td>
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<tr>
<td>Intra-mural and Inter-arterial</td>
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<td>N=26</td>
</tr>
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<td>Inter-arterial Only</td>
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## Primary Operations

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<th>Inter-arterial without intra-mural (N=8)</th>
<th>Not inter-arterial or intra-mural (N=1)</th>
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<td>CABG</td>
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<td>1</td>
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<tr>
<td><strong>Non-operative</strong></td>
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<tr>
<td><strong>Controls Available</strong></td>
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