Trends in surgical management of anomalous aortic origin of the coronary artery over 2 decades

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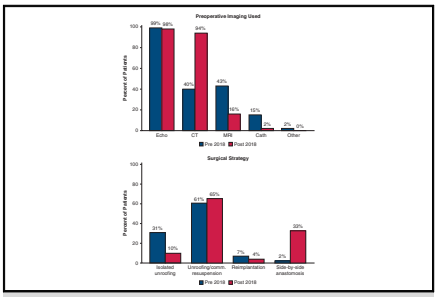
ABSTRACT

Objective: To evaluate outcomes of patients undergoing surgery for anomalous aortic origin of the coronary artery (AAOCA) at a tertiary care center and determine the influences of a coronary artery program on management strategies and outcomes.

Methods: This retrospective review of consecutive surgical patients who had isolated AAOCA at a tertiary care center between August 1, 1999, and October 31, 2022, compared patient characteristics, interventional timing, and surgical strategies before and after program inception in 2018. Comparisons between time periods and anatomical subgroups were performed using Fisher exact and Wilcoxon rank-sum tests.

Results: Of 149 surgical AAOCA patients, 102 (69%) had AAO of the right coronary artery. Compared with AAO of the left coronary artery (AAOLCA), AAO of the right coronary artery (AAORCA) was associated with greater athletic participation (intramural, varsity, and college-level) (74% vs 43%; $P < .001$) and preoperative functional imaging (72% vs 49%; $P = .01$), but were less likely to have ischemic changes on functional imaging (5% vs 23%; $P = .03$) or any postoperative complications (7% vs 19%; $P = .04$). Moderate or greater aortic insufficiency occurred postoperatively in 1 (1%) of AAORCA and 1 (3%) of AAOLCA patients. After the coronary artery program inception, there was an increase among patients with AAOCA undergoing preoperative computed tomography angiography (pre-2018: 39 out of 98 [40%] vs post-2018: 48 out of 51 [94%]; $P < .001$) and a decrease in isolated AAOCA unroofing procedures performed (30 [31%] vs 5 [10%]; $P = .004$).

Conclusions: Surgical management of AAOCA evolved over time, and can be achieved with low instance of postoperative aortic insufficiency. Establishment of a coronary artery program has streamlined care. (JTCVS Open 2023;16:757-70)



A coronary artery program offers streamlined imaging and surgical strategy changes.

CENTRAL MESSAGE

A programmatic approach to the surgical management of AAOCA allows for streamlined functional/anatomic imaging as well as changes in dominant surgical strategy.

PERSPECTIVE

Given the rarity of AAOCA and dearth of data on long-term outcomes, management of AAOCA is not standardized and is often driven by provider preference. Standardization of pre- and postoperative care may enable uniformity of care. Our results comparing the era before and after development of an AAOCA management algorithm highlights the importance of standardization of care in rare diseases.

Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital cardiac anomaly with an estimated prevalence of 0.01% to 2%[1] and believed to be the second most common cause of sudden cardiac death (SCD) in otherwise healthy individuals.[2-6] Anomalous aortic origin of the right coronary artery (AAORCA) from the opposite sinus of Valsalva is 4 to 5 times more common than anomalous aortic origin of the left coronary artery (AAOLCA) from the opposite sinus of Valsalva; however, SCD and symptoms of ischemia have been more commonly associated with the latter.[3,7,8] Many anatomical features of AAOCA, such as an intramural course, high

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Abbreviations and Acronyms

AAOCA = anomalous aortic origin of a coronary artery
AAOLCA = anomalous aortic origin of a left coronary artery
AAORCA = anomalous aortic origin of a right coronary artery
AI = aortic insufficiency
CAP = coronary artery program
CTA = computerized tomography angiography
ECMO = extracorporeal membrane oxygenation
ICC = intercoronary commissure
MRI = magnetic resonance imaging
SCD = sudden cardiac death

takeoff from the aorta, or slit-like coronary artery orifice, have been linked with increased risk of ischemia.[3]

Often, AAOCA is asymptomatic; and when present, symptoms may be nonspecific (nonanginal).[4] When diagnosed during screening for family history, clearance for athletic participation, or incidental finding during unrelated imaging, controversy exists on the best management strategy.[9] Currently, guidelines suggest that surgical repair should be used to mitigate the risk of sudden cardiac events in patients who have experienced them or to prevent lingering uncertainty related to the potential risk of SCD in patients who are asymptomatic (particularly those who are heavily involved in athletics).[4,10,11] Patient/family anxiety also play a large role in surgical decision making given that patients have led largely healthy lives up to their diagnosis, and the emotional influence of this unexpected life change certainly take a toll.[12]

The optimal strategies for management of AAOCA have significantly evolved over the past 2 decades and led to development of a coronary artery program (CAP) at our center in 2018. The purpose of this study was to better understand the surgical management and outcomes of AAOCA over the past 20 years at our center and to investigate the influence of the CAP on treatment strategies and outcomes.

MATERIALS AND METHODS

This was a retrospective review of consecutive patients who underwent surgical intervention for isolated AAOCA at a tertiary care center between August 1, 1999, and October 31, 2022. Patients with other significant complex intracardiac defects (besides patent ductus arteriosus, patent foramen ovale, or atrial septal defect) were excluded. The establishment of the CAP in January 2018 was used to stratify the dataset into 2 distinct eras. We collected information on baseline patient characteristics including age, sex, presence of major noncardiac abnormalities/syndrome/genetic abnormalities, presenting symptoms, preoperative testing, surgical intervention, and postoperative follow-up from our institutional databases and electronic medical records. The institutional review board at our center approved the study protocol and publication of data on April 16, 2021 (No. IRB-P00038559). Patient written consent for the publication of the study data was waived by the institutional review board because these data were collected as part of routine clinical care and we were not providing any research-related treatment or intervention to the patients. No individual was identified/tracked for the purpose of analysis or presentation following the review of medical records and department databases to obtain variables of interest.

Coronary Artery Program (CAP)

The CAP was established to standardize management of children and young adults with coronary anomalies both congenital (eg, AAOCA and coronary ostial atresia), and acquired (eg, Kawasaki disease, postcardiac surgery coronary obstruction, and familial hyperlipidemia-related coronary disease). A multidisciplinary group of pediatric cardiac surgeons, pediatric cardiologists, adult cardiologists, nurse practitioners, and database experts convene bimonthly for case review/discussion. This group includes practitioners with expertise in coronary disease, including catheterization, cross-sectional imaging, and functional imaging. A standardized center-specific protocol to guide preoperative imaging, perioperative care, and long-term follow-up in children and young adults with coronary anomalies, specifically AAOCA, was instituted with inception of the CAP (Online Data Supplement). This algorithm provides guidelines on imaging, management, and follow-up for AAOCA. Factors that influence decision making on surgery versus observation include symptoms of ischemia, age at diagnosis, type of AAOCA (left vs right), presence of intramural/interarterial course, and functional testing findings. The final decision is shared, including patient and parent preference. Patients for whom there is no immediate plan for surgery are periodically followed by their cardiologist, and depending on age, anatomy, and participation in competitive sports, undergo exercise stress imaging every 1 to 3 years. If they develop symptoms such as chest pain, palpitations, or fainting, they are counseled to cease exercise until further evaluation by their cardiologist, including provocative testing. Decision on additional imaging and management is shared after multidisciplinary case discussion. This protocol is dynamic, undergoes review, and is periodically updated based on the latest recommendations available in the literature.

Preoperative Testing

Data on preoperative testing included available echocardiograms, coronary computerized tomography angiogram (CTA), cardiac magnetic resonance imaging (MRI), cardiac catheterization, and various forms of functional testing (exercise stress test, stress echocardiogram, stress myocardial perfusion, stress positron emission tomography, or dobutamine stress MRI).

Surgical and Postoperative Data

Data on surgical technique, duration of cardiopulmonary bypass, myocardial ischemia time, and occurrence of minor/major adverse events (minor: pleural/pericardial effusion, pneumothorax, unplanned readmission < 30 days after surgical discharge; major: mediastinitis, unplanned cardiac reinterventions before discharge, need for extracorporeal membrane oxygenation [ECMO], or death) were collected.

Postoperative Follow-up

Follow-up data included readmissions, surgical or catheter-based reinterventions on the coronary artery or aortic valve, follow-up echocardiographs, cross-sectional imaging, cardiac catheterization, and functional testing information. The last available echocardiogram report at follow-up was used to determine patients' aortic insufficiency (AI) status.

Statistical Analysis

Categorical variables are summarized as frequencies and percentages, and compared between time periods and anatomical subgroups using

Fisher exact test. Continuous variables are summarized with medians and ranges, and compared using the Wilcoxon rank-sum test. Analyses were performed in Stata version 16 (StataCorp).

RESULTS

Between August 1999 and October 2022, 149 patients underwent surgery for AAOCA (Figure 1): 110 (74%) with AAORCA from left coronary sinus, 34 (23%) with AAOLCA from right coronary sinus, 8 (5%) with AAOCA (all with AAORCA) with both right and left intramural coronaries, 4 (3%) with AAOLCA from a noncoronary sinus, and 1 (1%) AAOLCA with an intraseptal/intramyocardial course. Note that some patients fall into more than 1 diagnosis category. Demographic/clinical characteristics of these patients are presented in Table 1. A total of 17 (11%) patients were diagnosed with a genetic syndrome, with genetic diagnosis (Table E1) being more frequently identified in the recent era (6 [6%] vs 11 [22%]; $P = .012$) (Table 2). Median age at surgery was 13 years (range, 63 days-49 years). There were 9 (6%) patients who underwent an operation during infancy/early childhood, who were frequently symptomatic (episodes of cyanosis, atypical chest pain, or multiple episodes of syncope). Of note, a 65-day-old patient presented after having an apparent life-threatening event of gasping, choking, and not breathing for 10 seconds. The indication for immediate surgery in this AAOLCA patient was concern for regional wall motion abnormality and decreased ventricular function on dobutamine stress echocardiogram. A second patient just younger than 5 years presented in cardiogenic shock and was emergently cannulated through the right neck for venoarterial ECMO. Catheterization on ECMO demonstrated AAOLCA that arose at the sinotubular junction of the left sinus of Valsalva close to the intercoronary commissure (ICC) and had a vertical intramural course.

Including the 2 patients described above, 8 (5%) total patients experienced aborted SCD, among whom 3 (38%) required preoperative mechanical circulatory support (Table 1). Other commonly presenting symptoms included exertional chest pain, dizziness, and syncope, with similar distribution of these symptoms for both forms of AAOCA. In general, more AAORCA patients reported participation

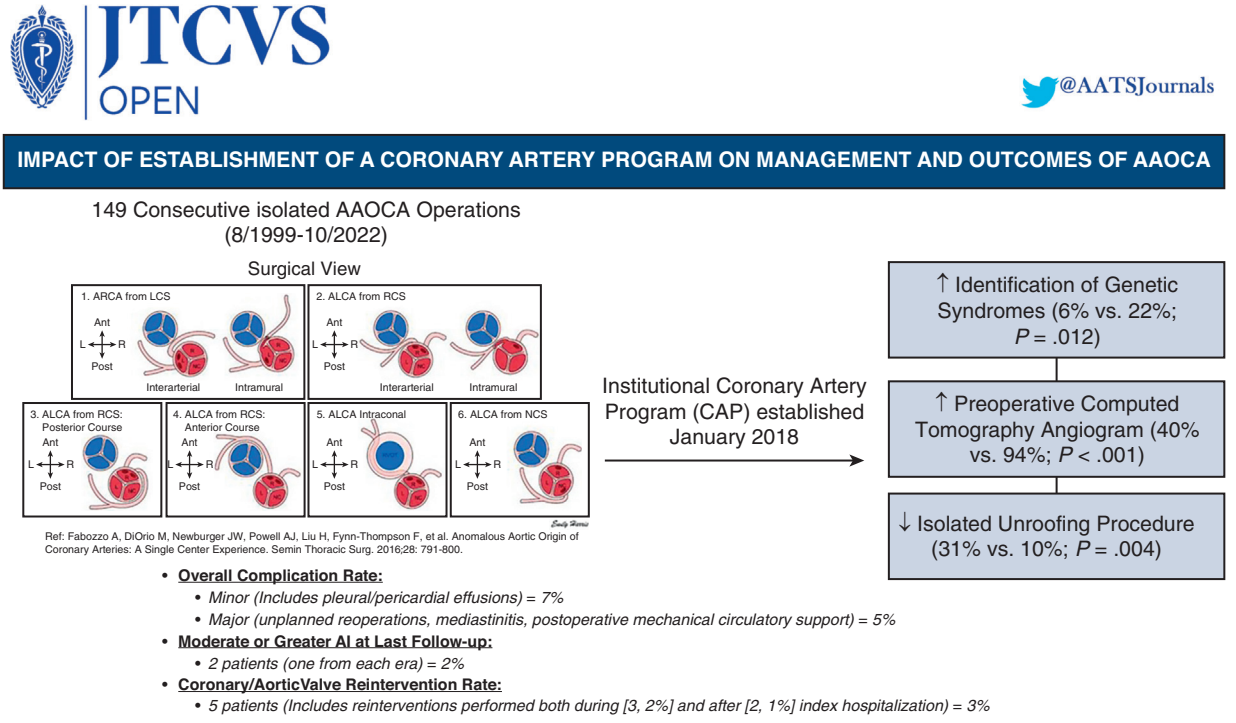

FIGURE 1. Impact of Establishment of a coronary artery program (CAP) on management and outcomes of anomalous aortic origin of a coronary artery (*AAOCA*). Beginning in 2018, the standardized protocols that were established as a part of this center's coronary artery program streamlined management of patients with AAOCA and aided in the clarity of surgical decision making. Our study emphasizes the importance of following an algorithm for management of this rare disease, given the dearth of long-term outcomes of its surgical repair. *LCS*, Left coronary sinus; *RCS*, right coronary sinus; *ALCA*, anomalous left coronary artery; *AI*, aortic insufficiency.

TABLE 1. Preoperative, surgical, and postoperative characteristics of the overall cohort of patients with anomalous aortic origin of a coronary artery (AAOCA) and in the anomalous aortic origin of right coronary artery (AAORCA) and anomalous aortic origin of left coronary artery (AAOLCA) subtypes

	Total cohort (N = 149)	AAORCA only (n = 102)	AAOLCA (n = 47)	*P* value
Age categories at AAOCA repair				
<5 y	9 (6)	4 (4)	5 (11)	
5-9 y	26 (17)	16 (16)	10 (21)	
10-19 y	103 (69)	74 (73)	29 (62)	
≥20 y	11 (7)	8 (8)	3 (6)	
Athletic activity	95 (64)	75 (74)	20 (43)	<.001
Associated intracardiac diagnosis	41 (28)	29 (28)	12 (26)	.84
Any symptoms	121 (81)	84 (82)	37 (79)	.65
If symptoms, age at onset (y)	12 (0.3-49)	12 (0.8-49)	12 (0.3-48)	.44
Echocardiogram performed	146 (98)	100 (98)	46 (98)	1.00
CT performed	87 (58)	64 (63)	23 (49)	.15
MRI performed	50 (34)	33 (32)	17 (36)	.71
Catheterization performed	16 (11)	11 (11)	5 (11)	1.00
Functional test performed	23 (15)	18 (18)	5 (11)	.34
Other diagnostic test	2 (1)	2 (2)	0 (0)	1.00
Preoperative echo	144 (97)	98 (96)	46 (98)	1.00
If yes, LVEF/LVFS in normal range (n = 96, n = 46)*	134 (94)	89 (93)	45 (98)	.44
Preoperative functional imaging	96 (64)	73 (72)	23 (49)	.010
If yes, type of test				.54
Stress echocardiogram	39 (41)	29 (40)	10 (43)	
Stress MIBI	32 (33)	23 (32)	9 (39)	
Stress PET	3 (3)	2 (3)	1 (4)	
Other: Exercise stress/stress MRI	22 (23)	19 (26)	3 (13)	
Symptoms at rest (n = 72, n = 23)	1 (1)	1 (1)	0 (0)	1.00
Symptoms at exercise (n = 72, n = 23)	23 (24)	18 (25)	5 (22)	1.00
Arrhythmia at rest (n = 72, n = 23)	12 (13)	9 (13)	3 (13)	1.00
Arrhythmia at exercise (n = 72, n = 23)	17 (18)	15 (21)	2 (9)	.23
Perfusion defect				.18
Normal/none	34 (35)	27 (37)	7 (30)	
Reversible	5 (5)	2 (3)	3 (13)	
No perfusion imaging	57 (59)	44 (60)	13 (57)	
Wall motion abnormalities† (n = 73, n = 22)	9 (9)	5 (7)	4 (18)	.21
Inducible Ischemic changes (n = 73, n = 22)	9 (9)	4 (5)	5 (23)	.029
Preoperative ECMO	3 (2)	0 (0)	3 (6)	.030
Surgical Timing				
Emergency surgery	3 (2)	0 (0)	3 (6)	.030
Planned surgery	146 (98)	102 (100)	44 (96)	.32
Reoperative AAOCA surgery‡	5 (3)	1 (1)	4 (9)	.035
Type of surgery				
Unroofing only	35 (23)	29 (28)	6 (13)	.039
Unroofing/comm. resuspension	93 (62)	59 (58)	34 (72)	.10
Reimplantation	9 (6)	3 (3)	6 (13)	.028
Side-by-side anastomosis	19 (13)	17 (17)	2 (4)	.037
Concomitant cardiac procedures	29 (19)	15 (15)	14 (27)	.085
CPB time	76 (41-249)	72 (41-161)	84 (46-249)	.30

(Continued)

TABLE 1. Continued

	Total cohort (N = 149)	AAORCA only (n = 102)	AAOLCA (n = 47)	*P* value
Crossclamp time	50 (21-173)	50 (22-109)	52 (21-173)	.24
Any adverse event	16 (11)	7 (7)	9 (19)	.043
Mediastinitis	1 (1)	1 (1)	0 (0)	1.00
Postoperative mechanical circulatory support	2 (1)	0 (0)	2 (4)	.098
Early surgical reintervention	3 (2)	1 (1)	2 (4)	.23
Late surgical reintervention	2 (1)	1 (1)	1 (2)	.53
Postoperative follow-up location				.21
Study center	91 (61)	66 (65)	25 (53)	
Outside facility	58 (39)	36 (35)	22 (47)	
Postdischarge complications (n = 89, n = 37)	10 (8)	6 (7)	4 (11)	.48
Postdischarge symptoms of ischemia (n = 88, n = 37)	4 (3)	4 (5)	0 (0)	.32
AI (n = 89, n = 38)				.035
None	94 (74)	72 (81)	22 (58)	
Trivial	21 (17)	11 (12)	10 (26)	
Mild	10 (8)	5 (6)	5 (13)	
Moderate	2 (2)	1 (1)	1 (3)	
Follow-up duration (y) (n = 100, n = 47)	1.6 (4 d-20.1 y)	1.6 (4 d-20.1 y)	1.7 (4 d-14 y)	.52

Values are presented as n (%) or median (range). *AAORCA*, Anomalous aortic origin of right coronary artery; *AAOLCA*, anomalous aortic origin of left coronary artery; *AAOCA*, anomalous aortic origin of a coronary artery; *CT*, computed tomography; *MRI*, magnetic resonance imaging; *LVEF*, left ventricular ejection fraction; *LVFS*, left ventricular fractional shortening; *MIBI,* myocardial perfusion imaging; *PET*, positron emission tomography; *ECMO*, extracorporeal membrane oxygenation; *Comm.*, commissural; *CPB*, cardiopulmonary bypass; *AI*, aortic insufficiency. *Numbers in parentheses represent the number of AAORCA then AAOLCA patients present in the dataset for that specific category. These numbers are only present when there was incomplete data available for that specific category. †Experienced at both rest and stress. ‡All 5 patients had initial surgery for AAOCA at an outside institution.

in athletic activity (intramural, varsity, or college-level) when compared with other forms of AAOCA (75 [74%] vs 20 [43%]; $P < .001$) (Table 1).

Diagnostic/Preoperative Testing

Table 3 provides information on commonly used diagnostic tests in this cohort. The most common functional imaging tests performed were stress echocardiogram and stress myocardial perfusion (Table 3). Compared with patients with AAOLCA, patients with AAORCA were more likely to receive preoperative functional imaging (73 [72%] vs 23 [49%]; $P = .01$) due to increased risk of SCD in those with an AAOLCA diagnosis. Those with AAOLCA who underwent preoperative functional testing were predominantly operated on in the pre-CAP era, and referred from another center where the functional imaging took place. Nevertheless, patients with AAORCA rarely (5%) exhibited ischemic changes on these tests compared with patients with AAOLCA (23%) (Table 1). When broken down by era, 94% of patients who received surgery post-2018 underwent a preoperative CTA, compared with just 40% in the pre-2018 era (Figure 2). Similarly, only 61% of patients who underwent surgery between 1999 to 2017 underwent preoperative functional imaging, with this number increasing to 71% in the 4 years since CAP establishment (Table 3). Of 96 (64%) patients across the study period who underwent preoperative functional imaging, only 9 (9%) patients exhibited inducible wall motion abnormalities or ischemic changes, despite 23 (24%) reporting chest pain during the test (Table 3).

Operative Technique

Characteristics of surgeries performed for AAOCA at this center are described in Table 4. Unroofing with commissural resuspension was performed in 93 patients (62%), with takedown of the commissure in 37 (25%). Isolated unroofing was performed in 35 patients (23%), side-by-side anastomosis/aortocoronary window in 19 patients (13%), and reimplantation in 9 patients (6%). Additional procedures such as patent foramen ovale or atrial septal defect closure, patent ductus arteriosus ligation, aortic patch plasty (in a patient who required additional unroofing and aortic valve plasty after prior incomplete unroofing was complicated by severe AI at an outside center), or insertable cardiomonitoring system placement were simultaneously performed in 29 patients (19%). Although the majority of procedures performed were elective and planned, 3 patients (2%), all with AAOLCA, required emergency surgeries.

TABLE 2. Baseline characteristics before and after establishment of the coronary artery program

	Total cohort (n = 149)	Surgery 1999-2017 (n = 98)	Surgery 2018-2022 (n = 51)	*P* value
Age at first presentation (y)	12 (10 d-49 y)	11 (15 d-49 y)	12 (10 d-46 y)	.92
Female	51 (34)	31 (32)	20 (39)	.37
Genetic abnormalities or syndromes	17 (11)	6 (6)	11 (22)	.012
Age at AAOCA diagnosis (y)	12 (5 d-49 y)	12 (16 d-49 y)	12 (5 d-46 y)	1.00
How patient reached center				.043
Primary patient	35 (23)	17 (17)	18 (35)	
Referred by outside cardiologist	111 (75)	79 (81)	32 (63)	
Self-referred	3 (2)	2 (2)	1 (2)	
AAOCA diagnosis before referral to our center	90 (60)	62 (63)	28 (55)	.38
AAORCA from LCS	110 (74)	72 (73)	38 (75)	1.00
AAOLCA from RCS	34 (23)	22 (22)	12 (24)	1.00
AAOCA with both IM coronaries	8 (5)	3 (3)	5 (10)	.12
ALCA intramyocardial/conal	1 (1)	1 (1)	0 (0)	1.00
AAOLCA from NCS	4 (3)	3 (3)	1 (2)	1.00

Values are presented as n (%) or median (range). *AAOCA*, Anomalous aortic origin of a coronary artery; *AAORCA*, anomalous aortic origin of right coronary artery; *LCS*, left coronary sinus; *AAOLCA*, anomalous aortic origin of left coronary artery; *RCS*, right coronary sinus; *IM*, intramural; *ALCA*, anomalous left coronary artery; *NCS*, non-coronary sinus.

Postoperative Course

There were no operative or late deaths. Patients with AAORCA were less likely to experience postoperative adverse events compared with other forms of AAOCA (7 [7%] vs 9 [19%]; $P = .043$). A total of 16 (11%) patients experienced minor (7%) and/or major (3%) postoperative adverse events (Table 4). The most common minor adverse events included pleural effusion (n = 4 [3%]), 1 of which required drainage; pericardial effusion requiring pericardiocentisis (n = 3 [2%]), and pneumothorax requiring drainage (n = 2 [1%]). One (5-year-old child) who initially presented in cardiogenic shock developed postoperative respiratory insufficiency that required mechanical ventilatory support for >7 days. Two (1%) patients were readmitted within 30 days of their AAOCA surgery. One presented to an outside facility emergency department complaining of chest pain. He was admitted overnight for observation and a CTA ruled out any coronary abnormality. The other was admitted for emesis and dehydration, and was treated with hydration therapy as well as ondansetron to alleviate symptoms of nausea before discharge.

Major adverse events included unplanned reoperation, re-exploration for bleeding, mediastinitis, central nervous system complications, ECMO, pacemaker, or mortality as defined by the Society of Thoracic Surgeons Congenital Heart Surgery Database. Three patients underwent unplanned reoperation during the index hospitalization for AAOCA repair. The first was a 14-year-old child who initially underwent isolated unroofing for AAORCA. The ICC was not taken down during initial unroofing, although the coronary was noted to run at the tip of the ICC. Postoperatively, he developed mild AI and right ventricular dysfunction with new onset of tricuspid regurgitation related to extrinsic right coronary artery compression that necessitated reoperation for lysis of tissue surrounding the right coronary artery. The AI was related to sagging of the tip of the ICC, which was therefore resuspended. The second patient was a 28-year-old adult with history of a coronary artery bypass graft with left internal thoracic to left anterior descending artery at an outside facility, with subsequent occlusion of the internal thoracic graft from competitive flow, and presented to our center with persistent intermittent chest pain. He underwent unroofing of an intraseptal AAOLCA at our center. Postoperative CTA raised concern for possible future development of compression of the left coronary artery, which was managed with insertion of a 30-mm main pulmonary artery interposition graft to relocate it more leftward. Finally, a 13-year-old patient with AAOLCA who underwent reimplantation was noted to have turbulent flow in the proximal left coronary artery on transesophageal echocardiography after cardiopulmonary bypass, and was immediately taken to the catheterization lab where the left coronary artery had a proximal narrowing of ~25%

TABLE 3. Diagnostic and preoperative testing before and after establishment of the coronary artery program

	Total cohort (N = 149)	Surgery before January 1, 2018 (n = 98)	Surgery after January 1, 2018 (n = 51)	*P* value
Associated intracardiac diagnosis	41 (28)	30 (31)	11 (22)	.33
Any symptoms	121 (81)	80 (82)	41 (80)	.83
If symptoms, age at onset (y)	12 (0.3-49)	12 (0.3-49)	12 (2-46)	.65
Competitive athletic activity/ interest	95 (64)	58 (59)	37 (72)	.15
Echocardiogram	146 (98)	97 (99)	49 (96)	.27
CT	87 (58)	39 (40)	48 (94)	<.001
MRI	50 (34)	42 (43)	8 (16)	.001
Catheterization	16 (11)	15 (15)	1 (2)	.012
Other diagnostic test	2 (1)	2 (2)	0 (0)	.55
Preoperative echo	144 (97)	94 (96)	50 (98)	.66
If yes, LVEF/LVFS in normal range (n = 134, n = 92, n = 50)	134 (94)	86 (93)	48 (96)	.71
Preoperative functional imaging	96 (64)	60 (61)*	36 (71)†	.28
If yes, type of test				<.001
Stress echocardiogram	39 (41)	18 (30)	21 (58)	
Stress MIBI	32 (33)	27 (45)	5 (14)	
Stress PET	3 (3)	0 (0)	3 (8)	
Other	22 (23)	15 (25)	7 (19)	
Symptoms at rest (n = 95, n = 59, n = 36)	1 (1)	1 (2)	0 (0)	1.00
Symptoms at exercise (n = 95, n = 59, n = 36)	23 (24)	14 (24)	9 (25)	1.00
Arrhythmia at rest (n = 95, n = 59, n = 36)	12 (13)	5 (8)	7 (19)	.20
Arrhythmia at exercise (n = 95, n = 59, n = 36)	17 (18)	13 (22)	4 (11)	.27
Perfusion defectd				.007
Normal/none	34 (35)	28 (47)	6 (17)	
Reversible	5 (5)	3 (5)	2 (6)	
No perfusion imaging	57 (59)	29 (48)	28 (78)	
Wall motion abnormalities‡ (n = 95, n = 59, n = 36)	9 (9)	6 (10)	3 (8)	1.00
Ischemic changes (n = 95, 59, 36)	9 (9)	7 (12)	2 (6)	.48

Values are presented as median (range) or n (%). *ECMO*, Extracorporeal membrane oxygenation. Values are presented as n (%) or median (range). *CT*, Computed tomography; *MRI*, magnetic resonance imaging; *LVEF*, left ventricular ejection fraction; *LVFS*, left ventricular fractional shortening; *MIBI*, myocardial perfusion imaging; *PET*, positron emission tomography. *Anomalous aortic origin of left coronary artery = 19%, anomalous aortic origin of right coronary artery = 81%. †Anomalous aortic origin of left coronary artery = 11%, anomalous aortic origin of right coronary artery = 89%. ‡Experienced at both rest and stress.

with a fractional flow reserve of 0.78 that necessitated revision with a patch plasty of the reimplanted coronary with a good result.

One patient required ECMO postoperatively. This was a 40-year-old woman with AAOLCA from the noncoronary sinus who initially underwent unroofing of the anomalous

TABLE 4. Surgical and perioperative data before and after establishment of the coronary artery program

	Total cohort (N = 149)	Surgery before January 1, 2018 (n = 98)	Surgery after January 2, 2018 (n = 51)	*P* value
Age at first surgery (y)	13 (63 d-49 y)	13 (63 d-49 y)	14 (3-47)	.24
Emergency surgery	3 (2)	1 (1)	2 (4)	.27
Planned surgery	148 (99)	97 (99)	51 (100)	1.00
Redo	5 (3)	2 (2)	3 (6)	.34
Preoperative ECMO	3 (2)	1 (1)	2 (4)	.27
Pump time (min) (n = 148, n = 97, n = 51)	76 (41-249)	65 (41-161)	93 (51-249)	<.001
Crossclamp time (min) (n = 148, n = 97, n = 51)	50 (21-173)	46 (21-109)	64 (41-173)	<.001
Unroofing only	35 (23)	30 (31)	5 (10)	.004
Unroofing and commissural resuspension	93 (62)	60 (61)	33 (65)	.72
Reimplant	9 (6)	7 (7)	2 (4)	.72
Side-by-side anastomosis	19 (13)	2 (2)	17 (33)	<.001
Additional procedure	29 (19)	15 (15)	14 (27)	.085
Any adverse event, minor and major*	16 (11)	8 (8)	8 (16)	.17
Mediastinitis	1 (1)	1 (1)	0 (0)	1.00
Postoperative mechanical circulatory support	2 (1)	1 (1)	1 (2)	1.00
Early surgical reintervention	3 (2)	2 (2)	1 (2)	1.00
Late surgical reintervention	2 (1)	2 (2)	0 (0)	.55

Values are presented as median (range) or n (%). *ECMO*, Extracorporeal membrane oxygenation. Values are presented as n (%) or median (range). *CT*, Computed tomography; *MRI*, magnetic resonance imaging; *LVEF*, left ventricular ejection fraction; *LVFS*, left ventricular fractional shortening; *MIBI*, myocardial perfusion imaging; *PET*, positron emission tomography. *Major adverse events are defined using Society of Thoracic Surgeons standards and include unplanned reoperation, re-exploration for bleeding, mediastinitis, central nervous system complications, ECMO, pacemaker, or mortality.

left main coronary artery into the left coronary sinus with commissural resuspension. Acute ischemic changes necessitated emergent cannulation to ECMO. Subsequent cardiac catheterization revealed acute spasm in both coronary systems managed with intracoronary infusion of nitroglycerin. The patient was decannulated without complications 2 days later.

Follow-up

The median duration of follow-up was 1.6 years (range, 4 days-20.1 years). In the current era, patients are more likely to follow up at our center rather than at an outside institution (Table 5).

Only 2 patients (2%) exhibited moderate or greater AI at their last follow-up (Figure 3). One had an extensive prior history, including 2 cardiac arrests and ECMO support for 5 days, and underwent initial left coronary artery unroofing with commissural resuspension at an outside institution. He subsequently required a tracheostomy and slide tracheoplasty for tracheal injury related to traumatic intubation. He was referred to our center for management of severe AI and was managed with additional left coronary artery unroofing and commissural resuspension. The second patient initially presented after a ventricular fibrillation arrest during athletic activity. He underwent unroofing and commissural resuspension for AAORCA and simultaneous implantation of a loop recorder that uncovered a tachycardia with a rate of 250. An electrophysiology study then successfully ablated a concealed left-sided pathway to retrograde conduction. His AI is being managed expectantly. Two patients who underwent initial surgery at our center required additional coronary surgery. One underwent AAOLCA unroofing without intercoronary commissure takedown at age 8 weeks for an acute life-threatening event and required completion of unroofing at age 8 years. The second patient had initial unroofing of AAORCA at age 10 months given several acute life threatening events, and subsequently at age 7 years underwent lysis of adhesions and unroofing of an intramyocardial left anterior descending artery due to persistent symptoms and the discovery of a myocardial bridge. Symptoms resolved following this operation.

About half (n = 82 [55%]) of this cohort underwent functional imaging during their postoperative follow-up. Of these, only 2 (2%), both patients with AAORCA,

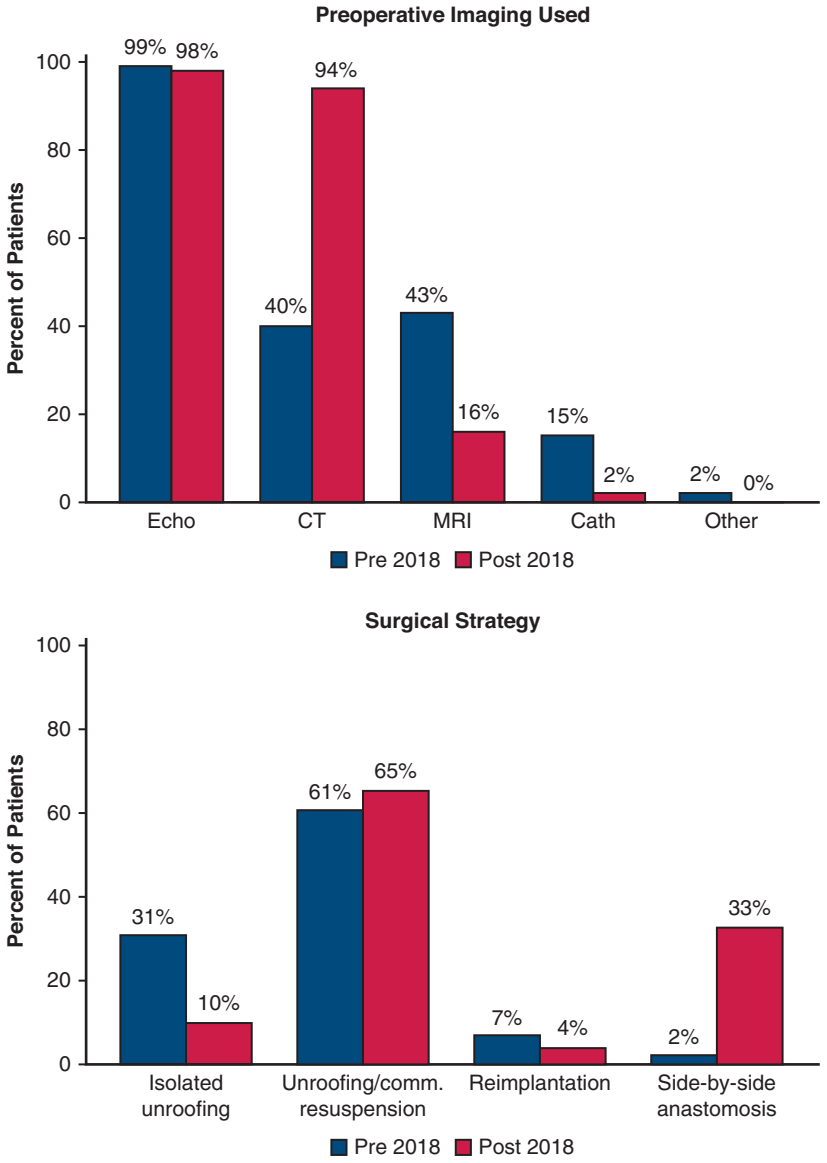

FIGURE 2. Surgical techniques and preoperative imaging by era distribution for anomalous aortic origin of a coronary artery repair during the 2 time periods of August 1, 1999-December 31, 2017, and January 1, 2018-October 31, 2022. There was a significant increase in the frequency of computed tomography (*CT*) angiography scans performed as well as the number of side-by-side anastomosis procedures performed following the establishment of the coronary artery program. *Echo*, Echocardiograph; *MRI*, magnetic resonance imaging; *Cath*, catheterization.

exhibited equivocal changes concerning for ischemia on functional tests (Table 5). The first patient's abnormal stress MIBI results were refuted on stress MRI. The second patient exhibited excellent exercise capacity on myocardial perfusion imaging with stress echocardiography; however, ischemia could not be excluded due to a small area of the inferior wall that was slightly thin and hypokinetic.

DISCUSSION

Our study illustrates the evolution of surgical management for patients diagnosed with AAOCA over the past 20 years at a single center. Given the risks associated with AAOCA, along with the variation in management strategies across centers,[3] many institutions have developed designated programs for coronary artery anomalies that evaluate/manage patients based on standardized algorithms.[2,4,9,12-17] The establishment of such a program at our center with protocols for preoperative testing, perioperative care, and long-term follow-up streamlined management in these patients and enabled us to compare the pre and post-CAP eras, thus highlighting the importance of standardization of care in rare diseases. Specific diagnostic testing modalities were performed with increased

TABLE 5. Follow-up data before and after establishment of the coronary artery program

	Total cohort (N = 149)	Surgery before January 1, 2018 (n = 98)	Surgery after January 1, 2018 (n = 51)	*P* value
Location				.008
Study center	91 (61)	52 (53)	39 (76)	
Outside facility	58 (39)	46 (47)	12 (24)	
Postdischarge complications (n = 126, n = 81, n = 45)	10 (8)	6 (7)	4 (9)	.74
Postdischarge symptoms of ischemia (n = 125, n = 81, n = 44)	4 (3)	3 (4)	1 (2)	1.00
AI (n = 127, n = 81, n = 46)				.27
None	94 (74)	57 (70)	37 (80)	
Trivial	21 (17)	14 (17)	7 (15)	
Mild	10 (8)	9 (11)	1 (2)	
Moderate	2 (2)	1 (1)	1 (2)	
Follow-up duration (y) (n = 147, n = 96, n = 51)	1.6 (4 d-20.1 y)	2.5 (4 d-20.1 y)	1.0 (4 d-4 y)	<.001
Follow-up functional imaging	82 (55)	53 (54)	29 (57)	.86
If yes, type of test				.036
Stress echocardiogram	55 (67)	30 (57)	25 (86)	
Stress MIBI	12 (15)	11 (21)	1 (3)	
Stress PET	2 (2)	2 (4)	0 (0)	
Other	13 (16)	10 (19)	3 (10)	
Symptoms at rest (n = 81, n = 52, n = 29)	2 (2)	2 (4)	0 (0)	.53
Symptoms at exercise (n = 81 n = 52, n = 29)	2 (2)	1 (2)	1 (3)	1.00
Arrhythmia at rest (n = 81, n = 52, n = 29)	20 (25)	13 (25)	7 (24)	1.00
Arrhythmia at exercise (n = 81, n = 52, n = 29)	9 (11)	5 (10)	4 (14)	.71
Perfusion defect				.008
Normal/none	15 (18)	14 (26)	1 (3)	
Fixed	1 (1)	1 (2)	0 (0)	
Reversible	2 (2)	2 (4)	0 (0)	
No perfusion imaging	64 (78)	36 (68)	28 (97)	
Wall motion abnormalities	11 (13)	8 (15)	3 (10)	.74
Ischemic changes (n = 81, n = 52, n = 29)	2 (2)	2 (4)	0 (0)	.53

Values are presented as n (%). *AI*, Aortic insufficiency; *MIBI*, myocardial perfusion imaging; *PET*, positron emission tomography.

consistency, and functional imaging was more selectively completed. Multidisciplinary case conferences allowed for informed decision making and ultimately shared decision making with patients/parents. This process resulted in a relatively low incidence of major adverse events (postoperative ECMO) of 1%, low reintervention rates on the coronary (2%) and aortic valves (2%), and moderate or greater AI rate of 2%, consistently lower than that reported in the multicenter Congenital Heart Surgeons' Society cohort, which included 395 surgical patients from 45 participating centers.[10] We believe the advantages to a coronary artery team is in standardization of care that surrounds surgical decision making, and steps that are taken to arrive at this important but often difficult decision.

The sole presence of AAOCA does not justify surgical correction, and therefore, thorough anatomic and

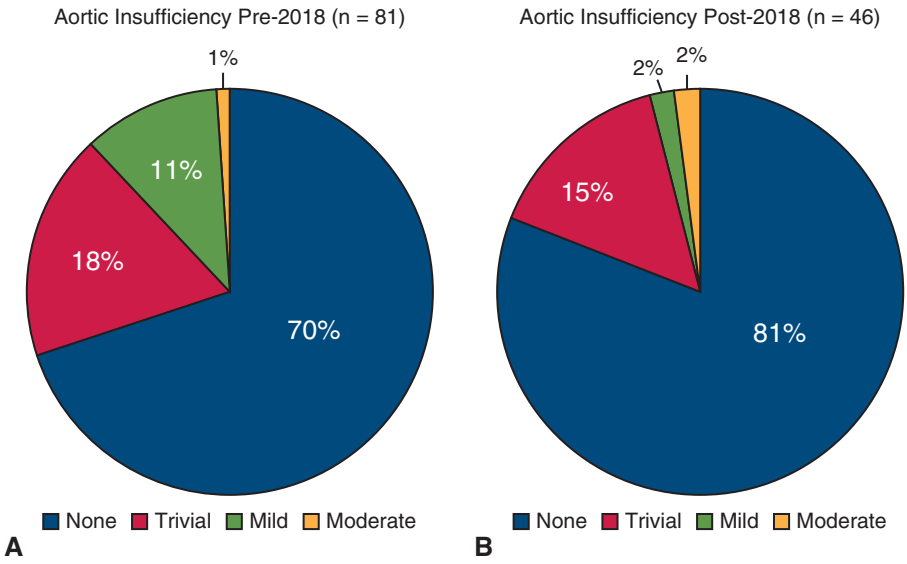

FIGURE 3. Incidence aortic insufficiency by era following anomalous aortic origin of a coronary artery repair for the 2 time periods of August 1, 1999-December 31, 2017 (A), and January 1, 2018-October 31, 2022 (B). There was a decrease in mild or greater aortic insufficiency in the post-2018 era.

hemodynamic assessment is needed initially.[18] Multimodality diagnostic testing should ideally be able to detect high-risk anatomic features of an AAOCA diagnosis; that is, an intramural/interarterial course, as well as myocardial ischemia.[18] Throughout our study period, transthoracic echocardiograms remain a mainstay in this pediatric population (98%). This is mainly due to this modality's ability to image the origin and proximal course of the coronary arteries without any radiation exposure.[18] Upon echocardiographic confirmation that AAOCA is present, it is important to highlight the paradigm shift in further testing preferences between the eras. Since CAP inception, CTA is the imaging modality of choice given its ability to delineate high-risk anatomic features (94%). Although MRI has the ability to assess the origin and proximal course of coronaries without radiation, and additionally provide information on myocardial scarring/viability; it often requires anesthesia in this pediatric population (as do catheterizations).[18] Therefore, CTA has proven to provide the best noninvasive spatial resolution to evaluate coronary anatomy.[18]

In our cohort, although patients with AAORCA are more likely than patients with AAOLCA to undergo preoperative functional testing, they are less likely to yield positive results. Many of these patients with AAORCA were first diagnosed because of exertional symptoms experienced during athletic participation that justifies the performance of a functional test preoperatively. In our cohort, we do not routinely perform provocative testing for AAOLCA with intra-arterial/intramural course, given the risk of SCD. Similarly, in the Congenital Heart Surgeons' Society cohort, 77% of patients who underwent preoperative functional testing had an AAORCA diagnosis. The optimal modality of functional testing for AAOCA is still unknown, and some patients who do in fact experience ischemia may be missed.[16] We observed a widespread variety of functional tests performed in this cohort, with stress echocardiograms becoming the preferred method in the current era. Conversely, other centers have used stress MRIs for functional testing given their feasibility/safety in the pediatric population as well as their increased accuracy compared with stress echocardiograms.[16] Given that stress MRI requires general anesthesia, particularly in younger patients, our center utilizes stress echocardiograms for initial provocative testing, with addition of a stress MRI only in patients with history suggestive of, or evidence of, inducible ischemia on other modalities such as exercise stress echocardiogram.

There are a number of common surgical techniques performed on patients diagnosed with AAOCA. Historically, unroofing procedures are most common.[3,4,10,17,18,19] Whereas a complete unroofing begins at the level of the ostium of the AAOCA, a partial unroofing starts anteriorly to the segment running behind the commissure. At our center, although a complete unroofing procedure with commissural resuspension remains the most common surgical technique, there has been a recent rise in the use of a side-by-side anastomosis/aortocoronary window in the post-CAP era. This strategy is completed by anastomosing the side of the coronary artery to a newly created ostia in the middle of the appropriate coronary sinus (depending on anatomy), and removes the need for

commissural manipulation completely. Thus, this strategy may decrease/avoid development of postoperative AI.[4,10] The 2 patients (1 transferred from another center after incomplete unroofing with takedown of ICC and resuspension) who exhibited moderate or greater AI on the most recent echocardiogram both underwent unroofing procedures with commissural resuspension. These trends were also observed in the Congenital Heart Surgeons' Society and other single-center cohorts, suggesting that surgical strategies avoiding commissural manipulation may decrease the risk of developing AI.[9,10,19,20] It should also be noted that since 2018, pump time and crossclamp time have increased. This is presumably due to this shift in surgical strategy preference, and although the short-term result is encouraging, a formal review of the side-by-side anastomosis technique is needed to determine this relatively novel approach's longevity. It is important to mention that trends seen in our experience, particularly those seen over the past 4 years since CAP inception, are to be considered with specific reference to our patient population, and a different center with different referral patterns and patient populations may have different findings.

Our study provides a larger experience with a population younger than age 10 years than many other recent reports.[2,13] Although we prefer to wait until a patient is at least 10 years old to operate, earlier surgery is indicated if a patient's quality of life is severely hindered, they are experiencing extreme symptoms, or if they have an AAOLCA diagnosis given higher risk of SCD in this cohort.

Limitations

Our study has the inherent limitations associated with a single center retrospective analysis, including bias from missing data and incomplete follow-up. This study covers 2 decades worth of data with changes in imaging and surgical techniques over time. Additionally, in the most recent era, which includes more than one-third of this study cohort, follow-up is limited to <3 years.

CONCLUSIONS

Surgical management of AAOCA has evolved over time, and can be achieved with low instance of postoperative AI and unplanned reintervention on the coronary or aortic valve. The establishment of a CAP streamlined care of these patients and allowed for standardized preoperative testing that aided in the clarity of surgical decision making. Following an algorithm for management in the short and long term is critical given the rarity of AAOCA, inability to clearly identify those at risk of SCD in AAORCA, and dearth of long-term outcomes of surgical repair of AAOCA. Multicenter registries such as the Congenital Heart Surgeons' Society AAOCA Registry, with ongoing long-term follow-up data collection, is key to answering questions that still remain.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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Key Words: anomalous aortic origin of a coronary artery, algorithm, aortic insufficiency, outcomes

TABLE E1. Genetic syndromes identified throughout study period

Genetic syndromes
Ellis-can Creveld syndrome
Goldenhar's syndrome
Trisomy 21
Turner syndrome
Beckwith-Wiedemann syndrome
Von Willebrand's disease
Ehlers Danlos syndrome (5 patients)
15 q. 11.2 microdeletion
Cloves syndrome
Hypochondroplasia
Epilepsy
Sickle cell disease
Unidentified malformation syndrome