Original Article

Congenital

Turk J Thorac Cardiovasc Surg 2026; 34(1): 46-55

Management of anomalous aortic origin of coronary arteries in children: A single-center experience

Mustafa Orhan Bulut[1], Gökçen Özçifçi[2], Onur Işık[3], Muhammet Akyüz[3], Tülay Demircan[4], Fatih Durak[5], Barış Güven[1]

[1]*Department of Pediatric Cardiology, University of Health Sciences Türkiye, İzmir City Hospital, İzmir, Türkiye*

[2]*Department of Pediatric Intensive Care, University of Health Sciences Türkiye, İzmir City Hospital, İzmir, Türkiye*

[3]*Department of Pediatric Cardiac Surgery, University of Health Sciences Türkiye, İzmir City Hospital, İzmir, Türkiye*

[4]*Department of Pediatric Cardiology, Dokuz Eylül University Faculty of Medicine, İzmir, Türkiye*

[5]*Department of Pediatric Intensive Care, İzmir Katip Çelebi University Faculty of Medicine, İzmir, Türkiye*

Abstract

Background: Anomalous aortic origin of coronary arteries (AAOCA) is the second leading cause of sudden cardiac death in young athletes and poses significant diagnostic and management challenges due to its variable anatomy and unpredictable clinical course.

Methods: We retrospectively analyzed 17 pediatric patients (10 male, 7 female; median age 12 years) diagnosed with AAOCA between 2016 and 2025. All patients underwent echocardiography and electrocardiogram-gated computed tomography angiography. Risk stratification considered interarterial course, intramural segment, and ostial morphology.

Results: Anomalous right coronary artery was most common (n=11, 64.7%), followed by anomalous left coronary artery (ALCA) (n=4, 23.5%) and anomalous circumflex arteries (n=3, 17.6%). Interarterial course was present in 12 patients (70.6%), and intramural course in 9 patients (52.9%). The main presenting symptoms were exercise-induced chest pain (n=10, 58.8%), syncope (n=8, 47.1%), dizziness (n=7, 41.2%), and palpitations (n=6, 35.3%). Syncope was predominantly observed in ALCA patients with interarterial course. Among 15 patients aged ≥8 years who underwent exercise stress testing, 6 (40%) showed inducible ischemia. Nine patients (52.9%) underwent surgical intervention, including unroofing (n=7), ostial reimplantation (n=1), and pulmonary artery translocation (n=1). The remaining eight patients (47.1%) were managed conservatively, six of whom received beta-blocker therapy.

Conclusion: Transthoracic echocardiography reliably identified AAOCA, showing excellent concordance with computed tomography and surgical findings. Standardized echocardiographic protocols are crucial, especially for children presenting with unexplained syncope or exertional symptoms, and can guide optimal management strategies.

Keywords: Anomalous aortic origin of coronary arteries, sudden cardiac death, unroofing.

In young individuals, unexpected cardiovascular death is most commonly caused by hypertrophic cardiomyopathy, followed by anomalous aortic origin of coronary arteries (AAOCA) arising from the opposite Valsalva sinus and coursing intramurally, making it the second most common cause of sudden cardiac death in this population.[1,2] Although the exact prevalence of AAOCA remains uncertain, studies using echocardiography, cross-sectional imaging, cardiac catheterization, and postmortem examinations suggest a range of 0.1-1% across different populations.[3,4] Most patients with AAOCA remain undiagnosed due to their asymptomatic nature.[1]

Transthoracic echocardiography (TTE) is the primary non-invasive modality for evaluating AAOCA in pediatric patients.[5-7] Its widespread availability, lack of radiation exposure, and absence of sedation requirements make it particularly suitable for children.

This study presents our single-center experience with 17 consecutive pediatric patients diagnosed with AAOCA, detailing their clinical evaluation, imaging findings, and surgical management.

Methods

This retrospective, single-center study was conducted following approval from Institutional Ethics Committee of University of Health Sciences Türkiye, İzmir Tepecik Education and Research Hospital (approval number: 29.04.2023/14-24, date: 23.12.2020). We systematically reviewed

Cite this article as: *Bulut MO, Özçifçi G, Işık O, Akyüz M, Demircan T, Durak F, et al. Management of anomalous aortic origin of coronary arteries in children: a single-center experience. Turk J Thorac Cardiovasc Surg. 2026; 34(1): 46-55*

Corresponding Author: *Barış Güven,*
e-mail: *drbarisguven@yahoo.com* ***ORCID:*** *orcid.org/0000-0002-4520-5574*
DOI: *10.4274/tjtcs.2025.27931*
Received: *May 22, 2025* ***Accepted:*** *Nobember 07, 2025* ***Publication Date:*** *January 08, 2026*

Copyright© 2026 The Author(s). Published by Galenos Publishing House on behalf of Turkish Society of Cardiovascular Surgery and Turkish Thoracic Surgery Association. This is an open access article under the Creative Commons AttributionNonCommercial 4.0 International (CC BY-NC 4.0) License.

departmental databases to identify pediatric patients diagnosed with AAOCA between January 2016 and April 2025.

The initial diagnostic evaluation for all patients included electrocardiogram (ECG) and TTE. During TTE, the following anatomical features were systematically assessed:

1. Location of the coronary ostium relative to the sinotubular junction (STJ);

2. Presence of interarterial and/or intramural course, including its length (Figure 1, Video 1);

3. Presence of an intraseptal or intraconal course of the proximal coronary artery.[8]

Patients with suspected coronary artery anomalies on echocardiography subsequently underwent ECG-gated computed tomography angiography (CTA) for detailed anatomical evaluation of coronary architecture.

Ischemia was assessed using treadmill exercise stress testing (EST) and/or ^{99}mTc-sestamibi stress nuclear perfusion imaging in patients older than eight years with CTA-confirmed AAOCA. EST was not performed prior to imaging confirmation due to the risk of exercise-induced ischemic events in patients with anomalous coronary anatomy.[9]

All patients underwent comprehensive risk stratification based on established anatomical criteria. High-risk features included interarterial course, intramural segment, slit-like ostium, or ostial stenosis. This stratification guided subsequent management decisions, particularly regarding the need for surgical intervention.

Surgical Techniques and Decision-making

Surgical intervention was indicated for symptomatic patients with high-risk anatomical features. The choice of technique was based on coronary anatomy:

1. Unroofing: Performed in patients with intramural coronary segments. The common wall between the aortic lumen and the intramural coronary artery was incised, with tacking sutures placed externally to prevent ostial narrowing.

2. Ostial reimplantation: Applied when the anomalous artery lacked an intramural course and arose from a separate ostium, involving anatomic relocation to the appropriate sinus.

3. Pulmonary artery (PA) translocation: Used when coronaries originated from a common ostium without an intramural course; the PA was laterally translocated to prevent coronary compression.

All procedures were performed via median sternotomy under cardiopulmonary bypass. The surgical technique was determined by preoperative imaging and intraoperative findings (Figures 2 and 3).

Excellent concordance was observed between anatomical details obtained from preoperative CTA and TTE and intraoperative findings, validating our diagnostic approach.

Statistical Analysis

Data were analyzed using SPSS version 24.0 (IBM Corp., Armonk, NY). Continuous variables were assessed for normality using the Shapiro-Wilk test. Normally distributed data are presented as mean ± standard deviation, while non-normally distributed data are expressed as median (range). Categorical variables are presented as frequency (n) and percentage (%).

Comparisons of categorical variables between surgical and conservative management groups were performed using Fisher's exact test. Effect sizes for proportional differences were calculated using Cohen's h, interpreted as small (h=0.2), medium (h=0.5), or large (h=0.8). Due to the small sample size (n=17), p-values were interpreted cautiously, with emphasis on effect sizes and clinical relevance. A two-tailed p-value <0.05 was considered statistically significant.

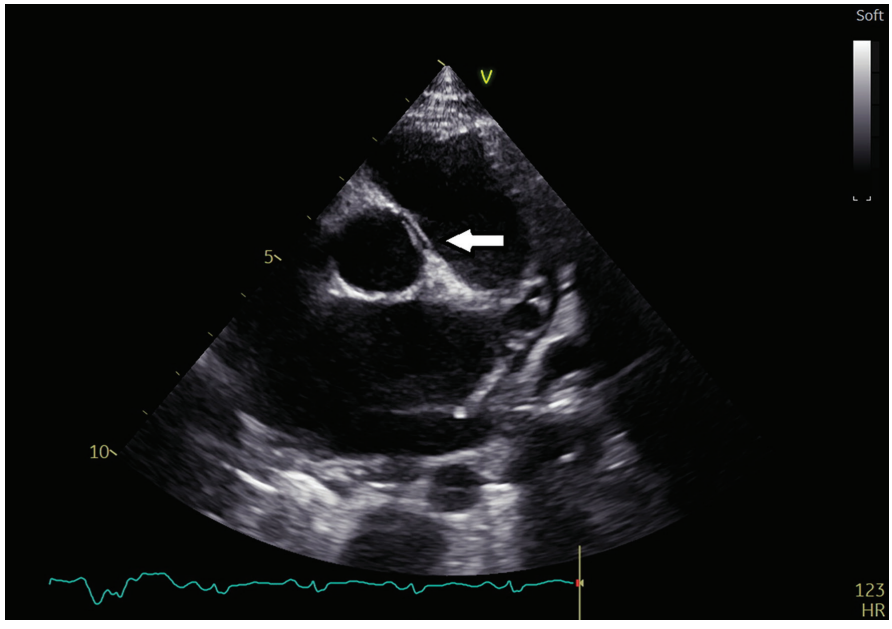


Figure 1. TTE parasternal short-axis view of Patient 14, demonstrating the intramural course of the anomalous left coronary artery. The white arrow indicates the intramural segment.

TTE: Transthoracic echocardiography.

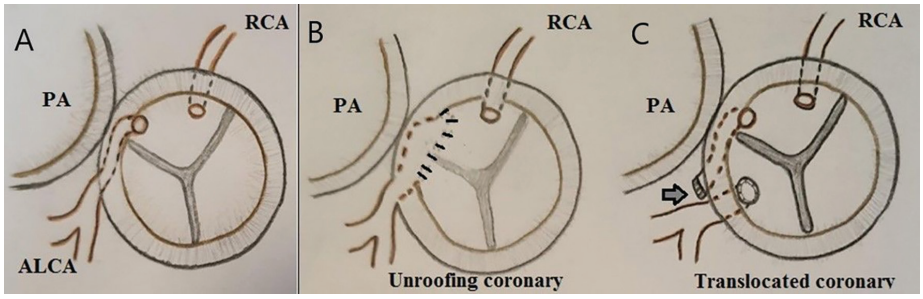


Figure 2. Surgical correction techniques for anomalous left coronary artery originating from the right coronary sinus: (A) Preoperative anatomy showing the left coronary artery arising from the right coronary sinus; (B) Unroofing procedure moving the ostium of the left coronary artery to the left coronary sinus; (C) Ostial translocation with re-anastomosis of the left coronary artery ostium into the left coronary sinus.

RCA: Right coronary artery; PA: Pulmonary artery; ALCA: Anomalous left coronary artery.

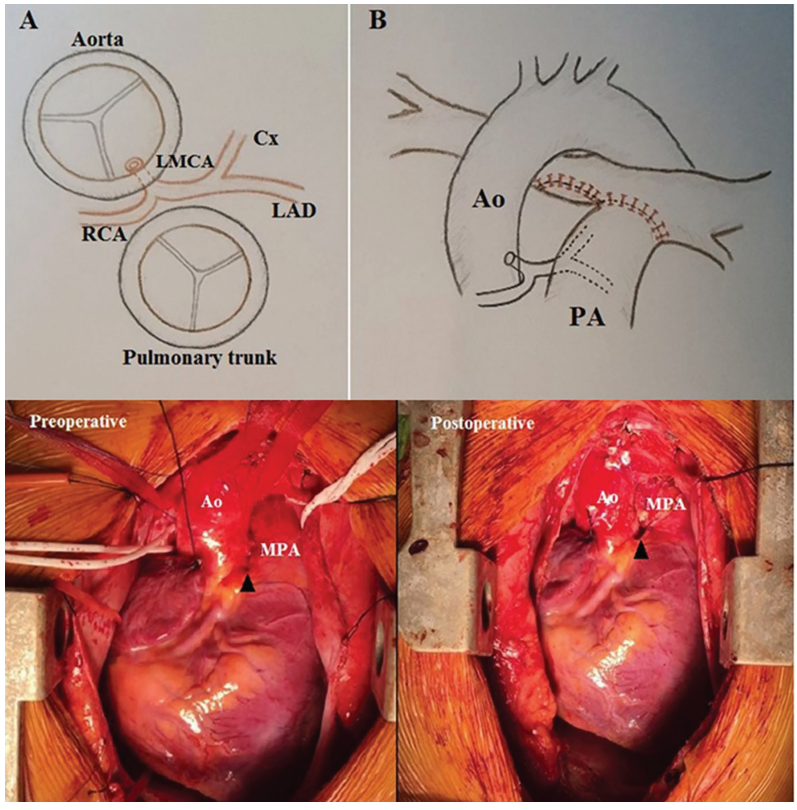


Figure 3. Anomalous left coronary artery originating from a common ostium in the right coronary sinus. (A) Axial view demonstrating the morphology of the abnormal origin. (B) Intraoperative view of PA lateral translocation, showing division of the main PA with re-anastomosis to the left PA. The black arrow indicates the increased distance between the aorta and PA.

Ao: Aorta; PA: Pulmonary artery; MPA: Main pulmonary artery; LMCA: Left main coronary artery; LAD: Left anterior descending artery; LCx: Left circumflex artery; RCA: Right coronary artery.

Results

Patient characteristics are summarized in Table 1. Seventeen pediatric patients (10 male, 7 female) with AAOCA were included. The median age was 12 years (range, 3-18 years), and mean weight was 42.7 kg (range, 17-70 kg). Clinical presentations were heterogeneous, with the most common symptoms being chest pain, syncope, dizziness, palpitations, and exertional angina. Notably, one patient presented with cardiac arrest requiring extracorporeal membrane oxygenation (ECMO).

The most frequent coronary anomaly was anomalous right coronary artery (ARCA), observed in 11 patients (64.7%), followed by anomalous left coronary artery (ALCA) in four patients (23.5%) and anomalous circumflex coronary artery (ACx) in three patients (17.6%). Interarterial course was present in 12 patients (70.6%), and intramural course in nine patients (52.9%). Among patients with an interarterial course, nine also had an intramural component, while three did not.

Nine patients (52.9%) underwent surgical correction, and eight (47.1%) were managed conservatively. All surgical patients had both interarterial and intramural courses and were symptomatic at presentation. In the surgical group, 100% (9/9) were symptomatic, compared with 62.5% (5/8) in the conservative group (Fisher's exact test, p=0.082; Cohen's h =1.32, indicating a large effect size). This substantial effect size highlights the

Table 1. Characteristics and clinical features of patients diagnosed with anomalous aortic origin of a coronary artery

Patient number	Age (y)	Sex	Weight (kg)	Symptoms	Anomaly	Course		Additional pathology	Treatment	Follow-up
						Intamural	Inter arterial			
1	12	M	49	Syncope, dizziness, exercise-related angina	ALCA	-	+	-	PA translocation	*13 months
2	18	F	61	Dizziness, palpitations, exercise-related angina	ARCA	-	+	-	Ostial reimplantation	*24 months
3	12	M	43	Syncope, dizziness, exercise-related angina	ARCA	+	+	-	Unroofing	*20 months
4	16	M	62	Syncope, exercise-related angina	ARCA	-	+	-	Unroofing	*14 months
5	14	M	55	Syncope, palpitations, exercise-related angina	ALCA, LCx	+	+	-	LAD unroofing	*6 months
6	3	F	17	N/A	ARCA	-	-	-	-	12 months
7	11	F	31	Non-specific CP^, dizziness, palpitgations	ARCA	-	-	-	B-blocker	30 months
8	12	M	35	Non-specific CP	ARCA	+	+	-	Unroofing	6 months
9	17	F	59	Non-specific CP, syncope, palpitations	ALCA	-	+	-	Refused surgery in our center** (coronary artery bypass graft at another center)	17 months
10	10	M	33	Non-specific CP, syncope, palpitations	LCx	-	-	-	B-blocker	15 months
11	5	M	19	Non-specific CP	ARCA	-	-	-	-	5 months
12	14	F	66	Non-specific CP	LCx	-	-	-	-	36 months
13	17	M	70	Non-specific CP, dizziness, exercise dyspnea	LCx	-	-	Renovascular HT	B-blocker ACE-I	15 months
14	8	F	20	Cardiac arrest ECMO syncope attacks at	ALCA	+	+	Systolic Dysfunction	Unroofing	6 months
15	12	M	34	Chest pain	ARCA	+	+		Follow-up^ sport restriction	5 months
16	7	F	23	Chest pain exercise	ARCA	+	+		Unroofing	3 months
17	12	M	45	Syncope attacks	ALCA	+	+		Unroofing	3 months

CP: Chest pain; TTE: Transthoracic echocardiography; ECG-CT: ECG-gated coronary computed tomography; N/A: Not applicable; HT: hypertension; B-blocker: Beta-blocker; ACE-I: Angiotensin-converting enzyme inhibitor; PA: Pulmonary artery.
*: Indicates the postoperative follow-up period of the patient; **: Patient underwent surgery at another center; ^: Patient 15: Surgery was declined by the family.

clinically meaningful difference in symptom burden between the two groups, reflecting the preferential selection of high-risk symptomatic patients for surgery.

The most common presenting symptoms were exercise-induced chest pain (n=10, 58.8%), syncope (n=8, 47.1%), dizziness (n=7, 41.2%), and palpitations (n=6, 35.3%). Syncope occurred in eight patients; in seven cases (41.2%), it was exertional, whereas one patient with an isolated ACx experienced a non-specific, non-exertional episode. One patient presented following cardiac arrest, requiring ECMO support. Three patients (17.6%) were asymptomatic, with anomalies detected incidentally during evaluation for unrelated conditions.

Resting ECG was normal in 11 patients (64.7%). Abnormal findings were observed in six patients (35.3%), including left ventricular hypertrophy in two, right bundle branch block in one, and isolated T-wave abnormalities in three.

Among 15 patients aged ≥8 years who underwent EST, 6 (40%) demonstrated inducible ischemia. ST-segment depression ≥1 mm

occurred in four patients, T-wave inversion in the precordial leads in three (Figure 4), and chest pain accompanied by dynamic ECG changes in five. Exercise-induced arrhythmias were documented in two patients (13.3%); both had premature ventricular contractions, and in one patient the arrhythmia met criteria for non-sustained ventricular tachycardia.

Surgical intervention was performed in nine patients (52.9%) at our center; all had both interarterial and intramural courses. One additional patient declined surgery at our center and underwent the procedure elsewhere. Conservative management was chosen for eight patients (47.1%). Of these, six received beta-blocker therapy, three were advised activity restriction, and two were managed with surveillance alone without pharmacologic treatment. The primary reasons for non-operative management were asymptomatic ARCA without high-risk anatomical features (n=5) and patient or family preference (n=2). One patient with renovascular hypertension received combined beta-blocker and angiotensin-converting enzyme inhibitor therapy.

Patient 15, a 14-year-old male with interarterial ARCA and an intramural segment, also had a slit-like ostium on CTA. He presented with exertional chest pain and ST-segment depression on exercise testing. Despite counseling regarding his high-risk anatomy, the family declined surgery. He was treated with metoprolol (25 mg twice daily), restricted from competitive sports, and followed at three-month intervals with ongoing counseling. Symptoms improved with therapy, and surgical intervention remains under reconsideration.

The mean follow-up period for all patients was 13.9±9.8 months (range, 3-36 months). Among surgically treated patients, mean postoperative follow-up was 11.3±7.2 months. Although the database review spanned 2016-2025, the standardized echocardiographic screening protocol for AAOCA was formally established in 2020. The 17 patients included in this study were diagnosed using this standardized protocol from 2020 onward.

The mean follow-up period was 13.9±9.8 months (range, 3-36 months), measured from diagnosis to the most recent clinical evaluation. Variability in follow-up duration was primarily due to patients diagnosed in 2020-2021 who were lost to follow-up or did not return for regular visits after initial treatment. No mortality was observed during the follow-up period.

All surgically treated patients demonstrated resolution of presenting symptoms, with no recurrence of syncope or exercise-induced chest pain during follow-up evaluations. Data for surgically treated patients are summarized in Table 2. Patient 9 was initially planned for unroofing but underwent coronary artery bypass grafting (CABG) of the left anterior descending and circumflex (CX) arteries at another center. The mean follow-up for patients operated on at our center was 15.9±6.9 months.

Unroofing was performed in seven patients due to the presence of an intramural course. During the procedure, the coronary ostium was evaluated with a right-angled clamp, and the length of the intramural segment was measured using a coronary probe. PA lateral translocation was performed in one patient, and coronary artery ostial translocation was performed in another.

Postoperative follow-up was conducted on the seventh day, first month, third month, sixth month, and annually thereafter. At each visit, patients underwent ECG and TTE evaluation. At the third-month follow-up, patients were specifically assessed for the development of symptoms during daily activities and high-intensity exercise (Tables 3 and 4).



Figure 4. Exercise ECG showing T-wave inversion in the precordial leads, indicative of ischemia during stress testing.
ECG: Electrocardiogram.

Table 2. Surgical group data

Patient no	Surgical indications	Ostial morphology	CPB time (min)	Cross clamp time (min)	Extubation time (hour)	Length of ICU stay (day)	Length of hospital stay (day)	Postoperative complication	Recurrent symptoms
1	ALCA, interarterial	Common ostium	118	61	4.5	2	5	N/A	N/A
2	ARCA, interarterial	Two separate ostia, slit like	126	68	8	1	4	N/A	Non-spesific chest pain
3	ARCA, intramural, interarterial	Common ostium	110	60	4	2.5	7	Pericarditis	N/A
4	ARCA, intramural, interarterial	Separate but adjacent ostia, slit like	96	49	4.5	2.7	7	Pericarditis	N/A
5	ALCA, intramural, interarterial LCx	LAD; separate ostia, LCx; common ostia with RCA	97	59	6	1	4	N/A	N/A
8	ARCA, intramural, interarterial	RCA separate ostium, RCA ostium slit like	95	61	5.5	2	5	N/A	N/A
14	ALCA intramural interarterial	LCA separate ostium	110	63	5	15	45	N/A	N/A
16	ARCA, intramural, interarterial	RCA separate ostium, RCA ostium slit like	90	32	4	1	4	N/A	N/A
17	ALCA intramural interarterial	LCA separate ostium	94	30	3.5	1	5	N/A	N/A

ALCA: Anomalous left coronary artery; ARCA: Anomalous right coronary artery; CPB: Cardiopulmonary bypass; ICU: Intensive care unit; LAD: Left anterior descending artery; LCx: Left circumflex artery; LCA: Left coronary artery; RCA: Right coronary artery, N/A: Not applicable.

Table 3. Surgical characteristics (n=9)

Variable	Value
Surgical technique, n (%)	
Unroofing	7 (77.8)
Ostial reimplantation	1 (11.1)
PA translocation	1 (11.1)
CPB time (min), mean ± SD	104.0±12.6
Cross-clamp time (min), mean ± SD	53.7±13.6
Extubation time (hr), median (range)	4.5 (3.5-8.0)
ICU stay (days), median (range)	2.0 (1.0-15.0)
Hospital stay (days), median (range)	5.0 (4-45)
Complications, n (%)	
Pericarditis	2 (22.2)
Mortality	0 (0.0)
Recurrent symptoms at follow-up	1 (11.1)†

†: Non-specific chest pain without evidence of ischemia; CPB: Cardiopulmonary bypass; ICU: Intensive care unit; SD: Standard deviation; PA: Pulmonary artery.

Discussion

Our findings highlight several important considerations regarding the clinical presentation, diagnostic evaluation, and surgical management of AAOCA in the pediatric population. TTE is essential for identifying this potentially fatal condition, as demonstrated in our cohort. AAOCA represents a surgically preventable cause of sudden cardiac death in young patients. In recent years, the number of diagnosed cases have increased significantly due to heightened awareness and improved detection.

Symptom presentation in AAOCA is highly variable, with a substantial proportion of patients remaining asymptomatic until a catastrophic event. While some patients experience palpitations, ventricular arrhythmias, presyncope, syncope, or exertional chest pain/angina, many present without warning signs.[9] This unpredictable clinical course underscores the importance of thorough TTE using standardized protocols, even in otherwise healthy individuals without cardiac symptoms. Following the identification of our first case, we implemented a standardized coronary artery visualization protocol during athletic screening examinations, adapted from established pediatric echocardiography practice standards,[8] as detailed in the methods section.

Table 4. Comparison between surgical and conservative groups

Variable	Overall (n=17)	Surgical group (n=9)	Conservative group (n=8)
Age (years), median (range)	12 (3-18)	12 (8-18)	11.5 (3-17)
Weight (kg), mean ± SD	42.7±16.4	47.9±15.8	36.9±16.2
Sex, n (%)			
Male	10 (58.8)	6 (66.7)	4 (50.0)
Female	7 (41.2)	3 (33.3)	4 (50.0)
Coronary anomaly, n (%)			
ARCA	11 (64.7)	6 (66.7)	5 (62.5)
ALCA	4 (23.5)	3 (33.3)	1 (12.5)
LCx	3 (17.6)	1 (11.1)	2 (25.0)
Anatomic features, n (%)			
Interarterial course	12 (70.6)	9 (100.0)	3 (37.5)
Intramural segment	9 (52.9)	9 (100.0)	0 (0.0)
Symptoms at presentation, n (%)			
Any symptom	14 (82.4)	9 (100.0)	5 (62.5)
Chest pain	10 (58.8)	6 (66.7)	4 (50.0)
Syncope	8 (47.0)	6 (66.7)	2 (25.0)
Dizziness	7 (41.2)	5 (55.6)	2 (25.0)
Palpitations	6 (35.3)	4 (44.4)	2 (25.0)
Ischemia on stress test, n/N (%)	6/15 (40.0)	5/9 (55.6)	1/6 (16.7)
Follow-up (months), mean ± SD	13.9±9.8	10.8±7.5*	17.4±11.5

*: Postoperative follow-up period for surgical patients; ARCA: Anomalous right coronary artery; ALCA: Anomalous left coronary artery; LCx: Left circumflex coronary artery; SD: Standard deviation.

Exertional symptoms such as chest pain, syncope, and palpitations were common in our cohort, reflecting myocardial ischemia during increased cardiac output. One patient presented with cardiac arrest requiring ECMO, emphasizing the life-threatening potential of AAOCA, particularly with ALCA, which carries a higher risk of sudden cardiac death than ARCA. Notably, 82.4% of patients in our series were symptomatic, contrasting with prior literature suggesting that AAOCA frequently remains silent until sudden cardiac death occurs.[10,11] This discrepancy likely reflects referral bias, as symptomatic patients are more commonly referred to tertiary cardiac centers for evaluation.

In our cohort, syncope was predominantly observed in ALCA patients (83.3% of syncope cases), differing from some published series. Molossi et al.[12] reported nearly identical syncope rates between ALCA (20%, 2/10) and ARCA (19.3%, 11/57). The interarterial course was present in all syncope cases in our series, consistent with Angelini et al.,[13] who identified this anatomical feature as strongly associated with clinical events. However, our overall syncope rate of 35.3% exceeds the approximately 20% reported by Finocchiaro et al.[14] and Molossi et al.,[12] suggesting either referral bias or heightened detection through our standardized diagnostic protocol. Clinically, Patient 17 experienced recurrent syncope and was diagnosed only on a second evaluation, highlighting the necessity of prompt echocardiographic interrogation of coronary origins in children and adolescents with unexplained syncope.

Our clinical experience aligns closely with the findings of Turner et al.,[15] demonstrating that TTE is an excellent first-line screening modality for AAOCA, particularly when performed by experienced sonographers. In their surgical series of 53 AAOCA cases, Turner et al.[15] reported that TTE accurately predicted the presence of an intramural course in 92.5% of patients. Similarly, our diagnostic pathway, which prioritizes TTE as the initial assessment, showed strong concordance with subsequent CT angiography findings and intraoperative anatomy.

While TTE is essential for initial screening, coronary computed tomography angiography (CCTA) has become indispensable for comprehensive risk stratification and surgical planning in AAOCA.[16,17] CCTA provides superior spatial resolution and allows quantitative evaluation of high-risk anatomical features that cannot be reliably assessed with echocardiography alone.[18] Specifically, CCTA accurately identifies intramural course length, ostial dimensions, proximal vessel narrowing, and coronary take-off angles—parameters that are prospectively correlated with hemodynamic significance and surgical outcomes.[19]

In our experience, measurements derived from CCTA directly informed surgical technique selection and contributed to excellent outcomes, with 100% of surgically treated patients experiencing resolution of presenting symptoms (Figure 5).

Our surgical approach is consistent with contemporary practice, with unroofing as the preferred procedure when an intramural segment is present. As described by Romp et al.,[20] this technique effectively eliminates the risk of coronary compression between the great vessels while establishing a reconstructed ostial implantation within the anatomically correct sinus. The absence of mortality and the excellent symptomatic improvement observed in our surgical cohort suggest that this approach is safe and effective in the pediatric population.

Surgical intervention was performed in 52.9% of our patients, reflecting the ongoing debate regarding the optimal management

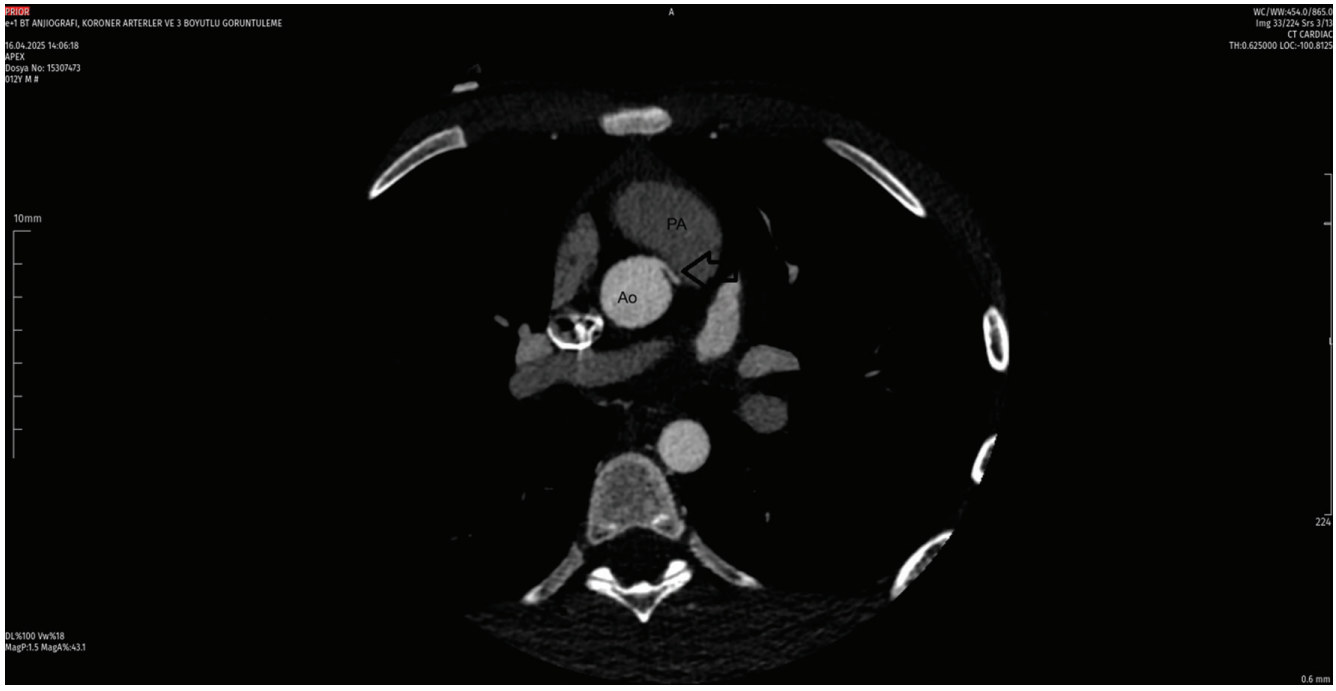


Figure 5. Computed tomography image showing anomalous origin of the left coronary artery (black arrow).
Ao: Aorta; PA: Pulmonary artery.

of asymptomatic AAOCA, particularly cases involving ARCA. Although conservative management is often advocated for asymptomatic ARCA, the 36th Bethesda Conference guidelines recommend restricting affected individuals from competitive sports—a limitation that may adversely affect quality of life in young athletes.[15] This consideration may influence decision-making toward surgical intervention, despite the low estimated risk of sudden death (~0.2% over 20 years).[11]

Although risk stratification for surgical treatment of AAOCA remains incomplete, recent advances have improved clinical decision-making. Molossi et al.[12] proposed a treatment algorithm in 2020 recommending surgical intervention for patients with long intramural courses, interarterial courses, or documented myocardial ischemia. Accordingly, all patients undergoing surgery in our cohort had a history of exertional angina, arrhythmia, or syncope. Among available surgical techniques, the most widely accepted methods include ostial excision and reimplantation, osteoplasty, PA translocation, and CABG in select cases, with unroofing remaining the approach of choice for patients with long intramural segments.

During unroofing, the common wall between the aortic lumen and the intramural coronary artery is incised to marsupialize the coronary, and tacking sutures are applied to ensure a widely patent neo-ostium. To minimize the risk of ostial narrowing due to endothelialization, suture knots are placed external to the aortic lumen.

When the anomalous artery lacks an intramural component and arises from a separate ostium, anatomic ostial reimplantation into the appropriate sinus is preferred over unroofing. For example, in an 18-year-old female patient, the RCA originated from the left coronary sinus with a separate ostium. The RCA was prepared as a button and reimplanted into the right coronary sinus below the STJ, and the aortic defect was closed with a glutaraldehyde-fixed pericardial patch. This patient has been followed for three years without ischemic symptoms.

When the right or left main coronary artery (LMCA) originates from a common ostium without an intramural course, PA lateral translocation is the preferred technique. This procedure prevents coronary compression between the great arteries by laterally repositioning the PA. For instance, in a 12-year-old male patient, the LMCA and RCA originated from a common ostium in the right coronary sinus. The LMCA divided into the LAD and CX branches after an interarterial course, and PA translocation was successfully performed.

Postoperative complications in our surgical cohort were minimal. Pericarditis was the only complication observed, which was promptly treated with ibuprofen, with no further sequelae. These findings are consistent with previously reported series.[17]

We did not perform CABG in any patient. Literature indicates that CABG in AAOCA is often associated with early graft occlusion due to competitive antegrade flow from the native coronary artery. Furthermore, anastomosis to an intact coronary vessel may cause vessel wall injury and subsequent narrowing. Fedoruk et al.[21] reported two ARCA cases treated with right internal mammary artery to RCA bypass at another institution; both patients experienced early graft failure and required reoperation—one underwent unroofing, and the other received coronary reimplantation.

After successful surgical repair, a gradual return to competitive sports is recommended once the patient is asymptomatic, ventricular function is normal, and maximal or symptom-limited exercise testing demonstrates no ischemia or arrhythmia. In the absence of residual high-risk anatomical features, clearance is typically considered at approximately three months postoperatively, individualized according to surgical findings and recovery.[22]

Advanced diagnostic modalities, including intravascular ultrasound (IVUS) and fractional flow reserve (FFR), show potential for AAOCA evaluation, although their routine clinical use remains investigational. IVUS enables

direct visualization of intramural segments and documentation of dynamic coronary compression throughout the cardiac cycle.[23] FFR provides functional assessment of stenosis severity and can identify hemodynamically significant lesions.[24] However, technical challenges in pediatric patients currently limit the widespread adoption of these invasive techniques.[25] Future studies should define the scenarios in which these modalities provide added value, particularly when non-invasive imaging is inconclusive or when functional data could alter management decisions.

Medical therapy is appropriate in cases of mild symptoms, absent or equivocal objective ischemia, or when surgery is deferred, typically in combination with activity modification. Beta-blockers are commonly prescribed to reduce heart rate and myocardial oxygen demand, potentially mitigating dynamic coronary compression. Calcium channel blockers may be considered when coronary vasospasm is suspected.[9,26]

Several limitations of our study merit consideration. The retrospective design and relatively small sample size (n=17) limit the generalizability of our findings and reduce statistical power, precluding multivariable analysis. Although comparative analysis revealed a large effect size for symptom burden between surgical and conservative groups (Cohen's h =1.32), the p-value (0.082) did not reach statistical significance, reflecting insufficient power rather than the absence of a clinically meaningful difference.

The short follow-up period (mean 13.9±9.8 months) restricts assessment of long-term outcomes. Selection bias is also inherent, as surgery was offered primarily to symptomatic patients with high-risk anatomical features. Despite these limitations, our findings contribute valuable insights into pediatric AAOCA and underscore the importance of individualized risk stratification.

Future research should focus on developing standardized risk stratification models capable of reliably predicting adverse outcomes in AAOCA. The Congenital Heart Surgeons' Society registry, initiated in 2009, represents an important step toward this goal.[27] Additionally, prospective studies comparing long-term outcomes of surgical versus conservative management in asymptomatic ARCA patients would provide critical guidance for clinical decision-making.

Collectively, these findings emphasize the necessity of prompt AAOCA diagnosis, appropriate selection of surgical techniques, and meticulous follow-up protocols to optimize outcomes in the pediatric population.

Video 1 Link: https://youtu.be/PTQoxZHQ1IM

Ethics

Ethics Committee Approval: This retrospective, single-center study was conducted following approval from Institutional Ethics Committee of University of Health Sciences Türkiye, İzmir Tepecik Education and Research Hospital (approval number: 29.04.2023/14-24, date: 23.12.2020).

Informed Consent: Retrospective study.

Footnotes

Authorship Contributions

Surgical and Medical Practices: G.Ö., O.I., F.D.; Concept: M.O.B., O.I., F.D.; Design: M.O.B., B.G.; Data Collection or Processing: G.Ö., M.A., T.D.; Analysis or Interpretation: G.Ö., M.A.; Literature Search: M.O.B., T.D., B.G.; Writing: M.O.B., B.G.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Jegatheeswaran A, Devlin PJ, McCrindle BW, Williams WG, Jacobs ML, Blackstone EH, et al. Features associated with myocardial ischemia in anomalous aortic origin of a coronary artery: a congenital heart surgeons' society study. J Thorac Cardiovasc Surg. 2019;158:822-34.e3. doi: 10.1016/j.jtcvs.2019.02.122.
2. Maron BJ, Friedman RA, Kligfield P, Levine BD, Viskin S, Chaitman BR, et al. Assessment of the 12-lead ECG as a screening test for detection of cardiovascular disease in healthy general populations of young people (12-25 years of age): a scientific statement from the American Heart Association and the American College of Cardiology. Circulation. 2014;130:1303-34. doi: 10.1161/CIR.0000000000000025.
3. Basso C, Maron BJ, Corrado D, Thiene G. Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. J Am Coll Cardiol. 2000;35:1493-501. doi: 10.1016/s0735-1097(00)00566-0.
4. Mery CM. Decision making in anomalous aortic origin of a coronary artery. Congenit Heart Dis. 2017;12:630-2. doi: 10.1111/chd.12493.
5. Frommelt PC, Berger S, Pelech AN, Bergstrom S, Williamson JG. Prospective identification of anomalous origin of left coronary artery from the right sinus of valsalva using transthoracic echocardiography: importance of color Doppler flow mapping. Pediatr Cardiol. 2001;22:327-32. doi: 10.1007/s002460010239.
6. Cohen MS, Herlong RJ, Silverman NH. Echocardiographic imaging of anomalous origin of the coronary arteries. Cardiol Young. 2010;20(Suppl 3):26-34. doi: 10.1017/S104795111000106X.
7. Walsh R, Nielsen JC, Ko HH, Sanz J, Srivastava S, Parness IA, et al. Imaging of congenital coronary artery anomalies. Pediatr Radiol. 2011;41:1526-35. doi: 10.1007/s00247-011-2256-3.
8. Lorber R, Srivastava S, Wilder TJ, McIntyre S, DeCampli WM, Williams WG, et al. Anomalous aortic origin of coronary arteries in the young: echocardiographic evaluation with surgical correlation. JACC Cardiovasc Imaging. 2015;8:1239-49. doi: 10.1016/j.jcmg.2015.04.027. Erratum in: JACC Cardiovasc Imaging. 2016;9:217.
9. Zhen Z, Dong Z, Na J, Chen X, Li Q, Gao L, et al. Clinical analysis of sixty-nine children with anomalous aortic origin of the coronary artery. Eur J Pediatr. 2023;182:4163-71. doi: 10.1007/s00431-023-05075-0.
10. Schubert SA, Kron IL. Surgical unroofing for anomalous aortic origin of coronary arteries. Operative Techniques in Thoracic and Cardiovascular Surgery. 2016;21:162-77. doi.org/10.1053/j.optechstcvs.2017.07.001
11. Brothers JA, Frommelt MA, Jaquiss RDB, Myerburg RJ, Fraser CD Jr, Tweddell JS. Expert consensus guidelines: anomalous aortic origin of a coronary artery. J Thorac Cardiovasc Surg. 2017;153:1440-57. doi: 10.1016/j.jtcvs.2016.06.066.
12. Molossi S, Agrawal H, Mery CM, Krishnamurthy R, Masand P, Sexson Tejtel SK, et al. Outcomes in anomalous aortic origin of a coronary artery following a prospective standardized approach. Circ Cardiovasc Interv. 2020;13:e008445. doi: 10.1161/CIRCINTERVENTIONS.119.008445.

13. Angelini P, Shah NR, Uribe CE, Cheong BY, Lenge V, Lopez JA, et al. Novel MRI–based screening protocol to identify adolescents at high risk of sudden cardiac death. JACC. 2013;61:E1621.

14. Finocchiaro G, Behr ER, Tanzarella G, Papadakis M, Malhotra A, Dhutia H, et al. Anomalous coronary artery origin and sudden cardiac death: clinical and pathological insights from a national pathology registry. JACC Clin Electrophysiol. 2019;5:516-22. doi: 10.1016/j.jacep.2018.11.015.

15. Turner II, Turek JW, Jaggers J, Herlong JR, Lawson DS, Lodge AJ. Anomalous aortic origin of a coronary artery: preoperative diagnosis and surgical planning. World J Pediatr Congenit Heart Surg. 2011;2:340-5. doi: 10.1177/2150135111406938.

16. Cheezum MK, Liberthson RR, Shah NR, Villines TC, O'Gara PT, Landzberg MJ, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of valsalva. J Am Coll Cardiol. 2017;69:1592-608. doi: 10.1016/j.jacc.2017.01.031.

17. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on clinical practice guidelines. J Am Coll Cardiol. 2019;73:e81-192. doi: 10.1016/j.jacc.2018.08.1029. Erratum in: J Am Coll Cardiol. 2019;73:2361-2. doi: 10.1016/j.jacc.2019.03.018.

18. Cantinotti M, Giordano R, Assanta N, Koestenberger M, Franchi E, Marchese P, et al. Echocardiographic screening of anomalous origin of coronary arteries in athletes with a focus on high take-off. Healthcare (Basel). 2021;9:231.

19. Farooqi KM, Nees SN, Smerling J, Senapathi SH, Lorenzoni R, Pavlicova M, et al. Assessment of anomalous coronary arteries by imagers and surgeons: comparison of imaging modalities. Ann Thorac Surg. 2021;111:672-81. doi: 10.1016/j.athoracsur.2020.03.124.

20. Romp RL, Herlong JR, Landolfo CK, Sanders SP, Miller CE, Ungerleider RM, et al. Outcome of unroofing procedure for repair of anomalous aortic origin of left or right coronary artery. Ann Thorac Surg. 2003;76:589-95; discussion 595-6. doi: 10.1016/s0003-4975(03)00436-3.

21. Fedoruk LM, Kern JA, Peeler BB, Kron IL. Anomalous origin of the right coronary artery: right internal thoracic artery to right coronary artery bypass is not the answer. J Thorac Cardiovasc Surg. 2007;133:456-60. doi: 10.1016/j.jtcvs.2006.10.011.

22. Padalino MA, Franchetti N, Hazekamp M, Sojak V, Carrel T, Frigiola A, et al. Surgery for anomalous aortic origin of coronary arteries: a multicentre study from the European Congenital Heart Surgeons Association†. Eur J Cardiothorac Surg. 2019;56:696-703. doi: 10.1093/ejcts/ezz080.

23. Formato GM, Agnifili ML, Arzuffi L, Rosato A, Ceserani V, Zuniga Olaya KG, et al. Morphological changes of anomalous coronary arteries from the aorta during the cardiac cycle assessed by IVUS in resting conditions. Circ Cardiovasc Interv. 2023;16:e012636. doi: 10.1161/CIRCINTERVENTIONS.122.012636.

24. Verheijen D, Egorova AD, Jongbloed MRM, Voskuil M, Van Der Kley F, Kies P, et al. Fractional flow reserve in anomalous aortic origin of coronary arteries to evaluate hemodynamic consequences of high risk anatomical features. European Heart Journal. 2023;44(Suppl 2). doi.org/10.1093/eurheartj/ehad655.1913

25. Agrawal H, Molossi S, Alam M, Sexson-Tejtel SK, Mery CM, McKenzie ED, et al. Anomalous coronary arteries and myocardial bridges: risk stratification in children using novel cardiac catheterization techniques. Pediatr Cardiol. 2017;38:624-30. doi: 10.1007/s00246-016-1559-4.

26. Rahmouni K, Bernier PL. Current management of anomalous aortic origin of a coronary artery: a pan-Canadian survey. World J Pediatr Congenit Heart Surg. 2021;12:387-93. doi: 10.1177/2150135121999030.

27. Poynter JA, Williams WG, McIntyre S, Brothers JA, Jacobs ML; Congenital Heart Surgeons Society AAOCA Working Group. Anomalous aortic origin of a coronary artery: a report from the Congenital Heart Surgeons Society Registry. World J Pediatr Congenit Heart Surg. 2014;5:22-30. doi: 10.1177/2150135113516984.