Surgical unroofing of intramural anomalous aortic origin of a coronary artery in pediatric patients: Single-center perspective

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ABSTRACT

Background: Intramural anomalous aortic origin of a coronary artery (AAOCA) is associated with an increased risk of sudden cardiac death. This is amenable to surgical coronary unroofing, but outcomes studies are lacking.

Objective: To perform a comprehensive review of our institutional experience with pediatric patients with AAOCA who underwent surgical repair with unroofing of the intramural segment, focusing on preoperative and postoperative course and testing as well as intraoperative findings.

Methods: A retrospective cohort study was conducted to evaluate patients with AAOCA status post-coronary unroofing at Children's Hospital of Wisconsin. Data extraction included symptoms, preoperative and postoperative imaging and testing, surgical findings, and postoperative clinical course.

Results: From January 1999 to December 12, 2015, 63 patients underwent unroofing at a median age of 13 years (0.5-18 years). The majority underwent unroofing of an intramural right coronary (79%); 21% had an intramural left AAOCA. Symptoms suggestive of possible ischemia were present in about 50%. Additional structural cardiac anomalies were present in 33%. Transthoracic echocardiography was diagnostic in 60 of 63 (95%) and correlated with surgical findings in all cases. There was no surgical mortality associated with the unroofing, and no additional coronary reinterventions were performed. The median duration of postoperative follow-up was 3.1 years (7 days to 13.6 years). Symptoms either persisted or developed in 46% postoperatively. Postoperative exercise stress testing, stress echocardiography, and cardiac magnetic resonance imaging were performed in 76%, 8%, and 20%, respectively, of the cohort. None identified findings consistent with reversible coronary ischemia. Three patients had sudden cardiac arrest (1 death) after surgery without an identified residual coronary abnormality.

Conclusions: Transthoracic echocardiography, with carefully designed coronary imaging protocols, can be diagnostic in accurately identifying intramural AAOCA in pediatric patients. Unroofing can be performed safely with no early morbidity, but symptoms can persist (including rare life-threatening events) without evidence of ischemia by postoperative provocative testing. (J Thorac Cardiovasc Surg 2018;155:1760-8)

Outcomes after unroofing of intramural anomalous aortic origin of a coronary artery (AAOCA). Shown are the postoperative outcomes in patients with AAOCA, status post unroofing surgery in terms of symptoms and investigation findings.

Central Message

Coronary unroofing can be performed safely, with minimal early morbidity, but symptoms can persist without evidence of ischemia by postoperative provocative testing.

Perspective

This study questions that unroofing is a curative procedure that relieves the risk of coronary ischemia in patients with intramural anomalous aortic origin of a coronary artery. This study highlights that, although surgery can be safely performed when the preoperative risk of myocardial ischemia is frequently unknown, risk stratification for subsequent events after unroofing is difficult to assess in the absence of standardized protocols.

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Isolated anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital heart defect (~0.2% of the population), but it is the second-leading cause of sudden

Scanning this QR code will take you to a supplemental video for the article.

Abbreviations and Acronyms

AAOCA	= anomalous aortic origin of a coronary artery
AAOLCA	= anomalous aortic origin of the left coronary artery
AAORCA	= anomalous aortic origin of the right coronary artery
AICD	= automatic implantable cardioverter-defibrillator
AR	= aortic regurgitation
CMR	= cardiac magnetic resonance imaging
CT	= computed tomography
ECG	= electrocardiogram
LV	= left ventricular
PVC	= premature ventricular contraction
SCA	= sudden cardiac arrest
TTE	= transthoracic echocardiogram

cardiac death in young athletes, after hypertrophic cardiomyopathy.¹ This anomaly appears to be of greater risk when the anomalous coronary courses intramural within the aortic wall before exiting the aorta from the contralateral sinus.² Children and adolescents can present with symptoms of myocardial ischemia, including exertional chest pain, syncope, or cardiac arrest.²⁻¹¹ Once identified, there is an ongoing controversy regarding exercise restriction, as the risk of acute myocardial ischemia is difficult to predict in the individual patient.¹²⁻¹⁴ Over the past 20 years, a surgical technique to address this pathology has been developed that involves unroofing of the intramural segment of the anomalous coronary artery (Video 1).^{7,15-21} Studies have shown improvement in symptoms postoperatively at experienced centers^{18,22-29}; however, there have been recent reports of late mortality after the unroofing procedure.³⁰ There is an ongoing controversy regarding exercise limitation and frequency and nature of follow-up studies in this population after surgery. Single-center studies have found conflicting results in assessing myocardial perfusion after coronary unroofing using stress echocardiography, exercise treadmill testing, and nuclear imaging, with discrepant findings between tests in the same patient.²³⁻²⁶

In this review, we provide a comprehensive description of our institutional experience in pediatric patients with intramural AAOCA who underwent surgical unroofing of the intramural segment, focusing on their preoperative, intraoperative, and postoperative course and testing.

METHODS

This was a retrospective cohort study of patients who underwent coronary unroofing surgery for an intramural AAOCA at Children's

Hospital of Wisconsin. This was approved by the Institutional Review Board (date and number of IRB approval: 8/25/2015, 782223-5). We identified all patients for this study by searching our institutional database from January 1999 (when coronary artery unroofing was first performed here) to December 2015. Medical record data were reviewed for demographic data, symptoms, preoperative investigations, surgical details, postoperative clinical course, and postoperative testing for the cohort. We reviewed the preoperative imaging to describe the anomalous coronary origin and course of the coronary artery as well as to identify other structural anomalies and assess ventricular function. All postoperative imaging was reviewed for coronary anatomy and course of the anomalous coronary, coronary flow patterns, ventricular wall motion abnormalities, and perfusion defects at rest and under pharmacologic stress if performed.

RESULTS

A total of 63 patients were identified who met the inclusion criteria. The patient characteristics are summarized in Table 1. The median age at diagnosis was 12.2 years (range, 1 day to 18 years). The median age at the time of surgery was 13 years (range, 0.5-18 years). Of the patients, 68% were male. The majority of our cohort (79%) had anomalous aortic origin of the right coronary artery (AAORCA), and 21% had an anomalous aortic origin of the left coronary artery (AAOLCA). Symptoms were present on presentation in 54% of the patients, with 44% having either an aborted sudden cardiac arrest (SCA) or exertional symptoms. The most common presenting symptom in both the groups was exertional syncope. There were 4 patients presenting after an aborted SCA event, of whom 3 had AAOLCA. Three of the patients (all with AAOLCA) presenting with an aborted SCA event had documented ventricular fibrillation during the event, and 1 patient had pulseless electrical activity; an additional

VIDEO 1. Unroofing procedure. This intraoperative video shows the unroofing procedure performed for intramural anomalous aortic origin of the right coronary artery from the left sinus with interspersed images detailing the procedure. The unroofing extended beyond the wall of the aorta, which required placement of sutures to reestablish continuity of the aortic wall to coronary artery. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(17\)32475-3/fulltext](http://www.jtcvsonline.org/article/S0022-5223(17)32475-3/fulltext).

TABLE 1. Patient characteristics

Patient characteristic	n (%)
Age at diagnosis	1 d to 18 y, median 12.2 y
Age at the time of coronary unroofing	6 mo to 18 y, median 13 y
Male	43 (68)
Anomalous right coronary	50 (79)
Anomalous left coronary	13 (21)
Intramural course	63 (100)
Symptomatic patients	34 (54)
Exertional chest pain	12
Nonexertional chest pain	5
Exertional syncope	16
Nonexertional syncope	2
Exertional dizziness	3
Exertional palpitations	1
Nonexertional palpitations	2
Aborted sudden cardiac arrest	4
Symptoms in anomalous right coronary	26/50 (52)
Exertional chest pain	10
Nonexertional chest pain	5
Exertional syncope	13
Nonexertional syncope	2
Exertional dizziness	3
Exertional palpitations	1
Nonexertional palpitations	2
Aborted sudden cardiac arrest	1
Symptoms in anomalous left coronary	8/13 (61)
Exertional chest pain	2
Exertional syncope	3
Aborted sudden cardiac arrest	3
Asymptomatic screening	29 (46)
Heart murmur	11
Detected during evaluation of known underlying cardiac or noncardiac diagnosis	16
Screening for Kawasaki disease	2
Intraoperative detection on TEE during surgery for other indications	1
Family history of coronary anomaly	1
Screening as a part of an unrelated research study	1
Additional cardiac diagnosis	21 (33)
Additional cardiac diagnosis associated with anomalous right coronary artery	18/50 (36)
Atrial septal defect	2
Bicuspid aortic valve	4
Coarctation of aorta	3
Double-outlet right ventricle with arch hypoplasia	1
Pulmonary atresia with intact ventricular septum and tricuspid stenosis	1
Pulmonary stenosis	2
Ventricular septal defect	7

(Continued)

TABLE 1. Continued

Patient characteristic	n (%)
Supraventricular tachycardia	1
WPW syndrome	1
Additional cardiac diagnosis associated with anomalous left coronary artery	3/13 (23)
Tetralogy of Fallot	1
Tricuspid atresia with pulmonary stenosis	1
WPW syndrome	1

TEE, Transesophageal echocardiogram; WPW, Wolfe-Parkinson-White.

patient with AAOLCA had documented nonsustained ventricular tachycardia when initially evaluated in the field after spontaneous recovery from the syncopal event. The 1 patient with aborted SCA and AAORCA had previous surgery for aortic coarctation and ventricular septal defect as an infant, and the coronary anomaly was only recognized after the SCA event at age 18 years. He presented with ventricular fibrillation during resuscitation and atrial arrhythmias after the SCA event with left ventricular dysfunction.

TABLE 2. Preoperative investigations

Preoperative investigations	n (%)
TTE	63 (100)
Diagnostic	60 (95)
Nondiagnostic	3 (5)
Site of coronary egress	
Opposite sinus	58/63 (92)
Posterior sinus	2 (3)
Ascending aorta	3 (5)
Ventricular function	
>55%	60 (95)
<30%	3 (5)
Cardiac CT	12 (19)
CMR	14 (22)
Wall motion abnormalities	0
Perfusion studies performed	5
Stress perfusion studies performed	4
Abnormal perfusion studies	0
Fibrosis/late gadolinium enhancement	0
Cardiac catheterization	8 (13)
ECG	51 (81)
Abnormal ECG	10/51 (19)
Holter test	3 (5)
Abnormal Holter test	0
Exercise treadmill stress test (Bruce)	27 (43)
Abnormal stress test	3/27 (5)
Stress echocardiogram	6 (10)
Abnormal stress echocardiogram	0
Troponin level	5 (8)

TTE, Transthoracic echocardiogram; CT, computed tomography; CMR, cardiac magnetic resonance imaging; ECG, electrocardiogram.

The remainder of the cohort (46%) was asymptomatic at the time of diagnosis, with incidental diagnosis made during echocardiographic evaluation of a heart murmur or ongoing echocardiographic evaluation of a known underlying cardiac or noncardiac diagnosis. An additional anatomic cardiac anomaly was found in 33% of all the patients (36% of patients with AAORCA and 23% of patients with AAOLCA), with ventricular septal defect and bicuspid aortic valve being the most common associated pathologies.

Preoperative Investigations

The preoperative investigations are summarized in Table 2. The most common site of egress for the anomalous coronaries was the opposite sinus (91.6% of the patients); the remainder were from the ascending aorta and posterior (noncoronary) sinus. All of the patients were identified preoperatively to have an intramural course with planned surgical unroofing except for 1 patient, who had the anomaly incidentally identified by intraoperative transesophageal echocardiography before surgery for other known congenital cardiac defects. Length of intramurality was not consistently assessed preoperatively and intraoperatively. Of the remaining 62 patients, 60 had the diagnosis made before surgery prospectively by transthoracic echocardiogram (TTE). TTE findings were confirmed by cardiac computed tomography (CT) in 12 patients, cardiac magnetic resonance imaging (CMR) in 14 patients, and cardiac catheterization in 8 patients; 36 patients went to surgery based on TTE alone. Two had the diagnosis made by cardiac CT (1 presented with an aborted SCA event who had limited TTE windows and the other presented with repeated exertional syncope and a suspected intramural coronary course by TTE; both had AAORCA). TTE identified 3 patients with an ejection fraction <30%; 2 had presented immediately after a cardiac arrest event, and 1 had tetralogy of Fallot that had been repaired without recognition of the intramural AAOCA (AAOLCA from the posterior sinus). All other patients had a normal ejection fraction before surgery.

There were no discrepancies in the coronary findings as assessed by other imaging investigations performed, including cardiac CT, CMR, or cardiac catheterization. Of the 14 patients who underwent CMR, 4 had stress perfusion studies. None of these studies showed any wall motion abnormalities, perfusion defects, or late gadolinium enhancement. Approximately 10% of the patients underwent a stress echocardiogram, and none of those studies was abnormal. None of the imaging studies could definitively identify coronary stenosis; intravascular ultrasound of the anomalous coronary was not performed during the cardiac catheterization procedure in any patient.

Eighty percent of the patients received a preoperative electrocardiogram (ECG); 19% were abnormal. These

TABLE 3. Intraoperative and postoperative course

Findings	n (%)
Duration of hospital stay	3-92 d, median 4 d
Single ostium, (1 AAOLCA, 4 AAORCA)	5 (8)
Ostial stenosis	21 (33)
Associated with AAORCA	18
Associated with AAOLCA	3
Immediate postoperative TEE	61 (97)
Patent unroofed coronary	61
Abnormal cardiac function	2
Discharge echocardiogram	63 (100)
Patent unroofed coronary	63 (100)
Abnormal cardiac function	2 (3)
EF 45%-55%	1
EF 30%-45%	1
Duration of follow-up	7 d to 13.6 y, median 3.1 y
Lost to follow-up	13/63 (21)
Died—sudden cardiac arrest	1/50 (2)
Resolution of symptoms	24/34 (71)
Persistent/new symptoms	23/50 (46)
Exertional chest pain	6
Nonexertional chest pain	10
Nonexertional palpitations	3
Nonexertional dizziness	2
Shortness of breath with exercise	1
Nonexertional syncope	1
Sudden cardiac arrest	3
Postoperative pacemaker/ICD placement	4/50 (8)

AAOLCA, Anomalous aortic origin of left coronary artery; AAORCA, anomalous aortic origin of right coronary artery; TEE, transesophageal echocardiogram; EF, ejection fraction; ICD, implantable cardioverter defibrillator.

abnormalities consisted of nonspecific ST changes in 3 patients, isolated uniform premature ventricular contractions (PVCs) in 2 patients, preexcitation in 2 patients, and 1 each showing left ventricular hypertrophy, right bundle branch block, sinus bradycardia, and prominent right-sided voltages with strain (in a patient who also had double-outlet right ventricle).

Forty-three percent of the patients had an exercise stress test (treadmill with modified Bruce protocol); 5% were abnormal. The abnormal findings included ST depression, T-wave inversion with activity, or frequent PVCs (known from baseline ECG) that suppressed at peak exercise. Both patients who had ST changes with exercise had normal perfusion studies (CMR with stress perfusion testing and stress echocardiography) before surgery.

Troponin level was performed in 5 patients, and all of them were elevated. Three were obtained status post-aborted SCA. Two of these patients had AAOLCA and 1 had AAORCA. Of the other 2 patients, 1 presented with exertional syncope and the other with exertional chest

pain. Both of these patients had AAOLCA and had presented to the emergency department with symptoms.

Intraoperative and Postoperative Course

Surgical findings correlated with the preoperative testing in all cases. All of the patients underwent only coronary unroofing, and there have been no other cardiac surgical procedures performed after the unroofing during follow-up for the cohort. Ostial stenosis was not a consistent finding in symptomatic patients. There were no early deaths. Of the 61 patients who had immediate postoperative transesophageal echocardiography and discharge echocardiogram, only 2 had depressed ventricular function, unchanged from the preoperative echocardiogram (Table 3).

Median duration of follow-up postcoronary unroofing was 3.1 years, with a range of 7 days to 13.6 years. Thirteen patients were lost to follow-up. All patients were allowed to exercise without restriction 3 months after unroofing, as none had a consistent pattern of ischemia by postoperative testing. Three patients had a SCA after unroofing, with 1 death as a result of the event. Two of the 3 had presented with an aborted SCA before the unroofing procedure. The death occurred 2 years after unroofing surgery in a patient with coarctation of the aorta and a perimembranous ventricular septal defect that were repaired in infancy. He was lost to follow-up and re-presented at 18 years of age with a witnessed SCA during which he was found to be in ventricular fibrillation. Subsequent work-up with TTE, cardiac CT, and cardiac catheterization demonstrated the previously unappreciated finding of AAORCA from the left sinus with an intramural course and left ventricular (LV) dysfunction; right ventricular function was qualitatively normal. An ECG revealed atrial flutter versus atrial fibrillation leading to an electrophysiology study preoperatively without evidence of high-grade AV block or any inducible arrhythmia that could have explained his cardiac arrest.

Postoperatively after unroofing, he had persistent ventricular dysfunction and underwent placement of a transvenous dual-chamber automatic implantable cardioverter-defibrillator (AICD) before discharge. As an outpatient, normalization of ventricular function was noted by serial TTE, as well as a normal stress test and CT angiogram showing patent coronaries, done for nonexertional chest pain. He had been doing well and was routinely followed with no evidence of atrial or ventricular arrhythmias. Two years after the unroofing procedure, he suddenly collapsed while he was being driven to work. Emergency medical services was not able to resuscitate him. Review of his AICD recordings demonstrated acute ventricular fibrillation that failed to respond to multiple shocks. He did not undergo postmortem examination.

The other 2 patients had aborted cardiac arrest postoperatively. One had a repeat aborted SCA event while he was taking a shower, 4 years after the unroofing surgery,

FIGURE 1. CMR findings. Presence of delayed enhancement in the inferolateral wall of the left ventricle (shown by the blue solid arrow) in a patient who presented with aborted sudden cardiac after unroofing surgery for anomalous aortic origin of left coronary artery. CMR, Cardiac magnetic resonance imaging.

with documented ventricular tachycardia during the event. This same patient had initially presented after an aborted cardiac arrest event with documented ventricular fibrillation. TTE on admission revealed an AAOLCA from the right sinus of Valsalva. The patient needed extracorporeal membrane oxygenation support for 4 days after the initial SCA event because of intractable arrhythmias and poor LV function. Serial postoperative echocardiograms showed persistent LV dyskinesia with endocardial echo-brightness consistent with endocardial scar. After the initial SCA event, he underwent postoperative cardiac catheterization with an electrophysiology study that demonstrated unobstructed coronary flow; however, there was inducible nonsustained ventricular tachycardia. He underwent AICD implantation before discharge, and an appropriate shock for the AICD terminated his second SCA event 4 years later.

The other patient initially presented with exertional syncope while swimming and was noted to have AAOLCA from the right sinus of Valsalva. He underwent surgical unroofing and had a normal postoperative TTE, ECG, and exercise stress test and so was cleared to return to competitive athletics. He subsequently presented with aborted SCA 6 years after the initial surgery when he collapsed while sprinting at track practice with documented ventricular fibrillation during resuscitation. He underwent cardiac catheterization that showed no coronary stenosis. He underwent a CMR that showed delayed enhancement in the basal and mid left ventricular inferolateral wall (Figure 1) consistent with focal scar. An AICD was placed before discharge. One other patient underwent AICD placement postoperatively who had presented with an aborted sudden death event and documented ventricular fibrillation on initial presentation in view of frequent PVCs and

TABLE 4. Postoperative testing

Postoperative investigations	n (%)
Postoperative echocardiogram performed	50/50 (100)
Patent unroofed coronary	50 (100)
Abnormal cardiac function	0
Aortic regurgitation	6/50 (12)
Trivial	3
Mild	2
Severe	1
Stress echocardiogram performed	4 (8)
Abnormal stress echocardiogram	0
CMR	10/50 (20)
Wall motion abnormalities	0
Perfusion studies performed	10
Stress perfusion studies performed	8
Abnormal perfusion studies	1
Fibrosis/late enhancement	1
Cardiac catheterization performed	3/50 (6)
ECG performed	50/50 (100)
Abnormal ECG	8
Holter test performed	12/50 (24)
Abnormal Holter test	2
Stress test performed	38/50 (76)
Abnormal stress test	6

CMR, Cardiac magnetic resonance imaging; ECG, electrocardiogram.

ventricular couplets at peak exercise on his postoperative follow-up exercise stress test.

Of the remaining patients, 23 of 50 (46%) had new/persistent symptoms, 9 of which were asymptomatic preoperatively. A total of 6 of 12 (50%) patients with AAOLCA and 17 of 38 (44.7%) patients with AAORCA had persistent or new symptoms post unroofing surgery. Most common symptoms were nonexertional and exertional chest pain. Besides the 3 late SCA patients, none of the symptomatic patients had evidence of ventricular dysfunction or coronary ischemia by postoperative testing.

Postoperative Testing

All of the patients underwent TTEs postoperatively (Table 4), and all showed widely patent coronary origins with laminar flow by color Doppler. Postoperatively trivial aortic regurgitation (AR) was seen in 3 patients, and mild AR was seen in 3. Of the patients with mild AR, 1 had it preoperatively associated with a bicuspid aortic valve, and 2 developed it postoperatively. One patient continued to have chronic mild AR without change, whereas the other progressed to severe AR over time, requiring aortic valve repair 6 months after the unroofing surgery. Of note, this patient also had aortic commissural resuspension as part of the unroofing procedure. The other 4 patients who had aortic commissural resuspension as part of their unroofing procedure had no AR at follow-up.

Postoperative stress echocardiograms were performed in 4 patients, and all were normal with no inducible wall motion abnormalities. Postoperative CMR was performed in 10 patients. None had any ventricular wall motion abnormalities, and only 1 had a myocardial scar at rest in the basal and mid left ventricular inferolateral wall (AAOLCA with postoperative SCA, described in the Intraoperative and Postoperative Course section). The majority (8/10) had pharmacologic stress perfusion performed as part of the CMR, and no perfusion abnormalities were noted. Coronary imaging by CMR identified a diffusely small right coronary artery in 1 patient; this was also noted on cardiac catheterization without evidence of discrete coronary stenosis. One patient had global delayed enhancement after tetralogy of Fallot repair; he eventually underwent heart transplant for chronic severely depressed LV function that was also present before the unroofing.

Three patients had a cardiac catheterization performed postoperatively. Of these, 1 demonstrated a diffusely hypoplastic proximal segment of the unroofed RCA as described previously.

All of the patients underwent an ECG postoperatively. Six patients with abnormal postoperative ECGs had nonspecific ST changes, and 2 had isolated PVCs. Holter testing was performed in 12 patients; 2 were abnormal with frequent PVCs. One of these patients had identical ectopy preoperatively that suppressed with exercise. Thirty-eight patients underwent stress tests (76%) postoperatively; 6 were abnormal (nonspecific T-wave changes or isolated PVCs). PVCs were not suppressible with exercise in 2 of the patients; 1 had initially presented with aborted SCA with documented ventricular fibrillation but otherwise remained asymptomatic after initial recovery. He had AAOLCA with laminar flow in the unroofed coronary with normal function by postoperative TTE. He also had both a preoperative and a postoperative CMR that did not reveal any ventricular wall motion abnormalities, perfusion defects at rest and under pharmacological stress, or any areas of delayed enhancement. The other patient with PVCs not suppressed with exercise presented with exertional syncope initially and then presented 6 years after unroofing with an aborted SCA (as described previously). His postoperative CMR did show an area of delayed enhancement at rest. He did not have a preoperative CMR.

DISCUSSION

In this review, we have described our single-institutional experience with surgical unroofing of an intramural AAOCA in children and adolescents. Our cohort is one of the largest reported, and the description of postoperative findings is unique in providing median-term outcomes with this relatively new surgical technique in pediatric patients. The majority of our cohort had AAORCA,

approximately one half were symptomatic preoperatively, and the median age at surgery was the early teenage years, with no early surgical mortality. These findings are similar to the reports from the retrospective data review from the AAOCA CHSS registry involving 23 institutions nationwide.^{27,28} TTE reliably and prospectively diagnosed AAOCA in more than 95% of the cohort, and the echo findings were always consistent with the surgical descriptions of the anatomy. This degree of accuracy is inconsistent with the recent report by Lorber and colleagues,³¹ which found variable agreement between the TTE and surgical findings. We have had a focused echocardiographic protocol in identifying intramural AAOCA that has been crucial in our diagnostic success, with specific features always characterized: (1) abnormal ostial origin from the wrong sinus, frequently with a more superior ostial take-off; (2) acute angle of origin of anomalous coronary with a tangential rather than a perpendicular course from the aortic root; (3) a proximal interarterial coronary course within the aortic wall; and (4) identification of linear diastolic color flow in the proximal coronary as it courses within the aortic wall.^{18,22,32,33}

Although CMR is also being used as a reliable modality for diagnosis of these anomalies, no other abnormal findings, other than confirmation of the AAOCA, were noted in the 14 patients who underwent a preoperative CMR (with 4 also undergoing CMR stress perfusion testing) in our cohort.^{18,34-41} Other imaging techniques that have been described to characterize the proximal course and shape of the intramural coronary in adults, including CT angiography⁴² and intravascular ultrasound,⁴³ were not used in our cohort.

There was rapid recovery from the unroofing surgery with no significant early postoperative morbidity or mortality. The median duration of hospital stay was short at 4 days, and resolution of symptoms was noted in more than 70% of the symptomatic patients. TTE, in addition to being used reliably for diagnosis preoperatively, was also used consistently for postoperative assessment. It demonstrated anatomic patency and laminar antegrade flow in the proximal segment of unroofed coronary in all the patients with normal systolic function after unroofing surgery in those patients who had normal function preoperatively. Only 2 patients demonstrated new AR, with progression in one who had aortic valve resuspension as a part of the unroofing surgery and eventually needed aortic valve replacement. We have modified our technique after that case to only unroof the intramural segment that is within the appropriate sinus (so that the segment behind the commissure is not manipulated, and no re-suspension is required).¹⁵

Despite these reassuring findings, almost one half of the cohort either had persistence of symptoms or developed new symptoms postoperatively that could be suggestive of

FIGURE 2. Surgical trends in the number of intramural AAORCA (blue line) and intramural AAOLCA (red line) unroofing surgeries performed per year at Children's Hospital of Wisconsin. AAORCA, Anomalous aortic origin of right coronary artery; AAOLCA, anomalous aortic origin of left coronary artery.

cardiac ischemia. Follow-up studies after surgical unroofing to assess myocardial perfusion (both at rest and with exercise) and the reconfigured coronary anatomy are scant in this population after surgery. It is likely that many of the children who had symptoms of chest pain before and after surgery had a noncardiac etiology, especially given the lack of correlating positive provocative testing. This is a critical question because any compromise to coronary blood flow related to this surgery could significantly increase the risk for coronary ischemic events later in life.^{44,45} There is an ongoing controversy regarding the need for exercise limitations; guidelines from 2005 recommended strict activity restriction for all interarterial anomalous coronaries (and led to our recommendation that unroofing was reasonable to allow competitive sports).¹² New guidelines were recently published that allowed competitive athletics for a child or adolescent with an interarterial AAORCA if there was no evidence of ischemia.¹³ This change, in concert with the recognition that intramural AAORCA appears less prone to SCD in the pediatric age group, has led to a significant decline in surgical unroofing here (Figure 2).

In addition, the frequency and nature of follow-up studies in this population to predict subsequent events and therefore allow competitive activities remains unclear.^{24,46} Previous studies have shown subclinical changes suggestive of ischemia might occur after AAOCA unroofing surgery despite patent neocoronary ostia, notably after AAORCA repair.⁴⁶ It is also hypothesized that myocardial scarring, incomplete unroofing, and coronary distortion with unroofing may contribute to postoperative symptoms.

Importantly, significant events did occur postoperatively. There were 3 SCAs, including 1 death, of whom 2 patients

had initially presented after an aborted SCA. None of these patients had postoperative ischemic symptoms, but all showed evidence of myocardial injury (either at presentation with SCA and abnormal function or by late fibrotic changes by echo or CMR). Postoperative testing for the rest of the cohort did not identify any significant reversible ischemic changes. There was considerable variation among providers at our institution regarding postoperative follow-up investigations and exercise restriction. Also, different tests occasionally showed conflicting results in the same patient. Symptoms suggestive of cardiac ischemia either persisted or developed in many patients postoperatively, but there was no clear evidence of ongoing myocardial ischemia based on provocative testing (Central Figure). These findings were similar to what was noted in a previous study by Brothers and colleagues,⁴⁶ where only 1 symptomatic patient was noted to have an abnormal preoperative stress test, and postoperative testing showed variable and incongruent findings that did not clearly identify patients at continued risk.

This study has important limitations that include its retrospective nature, with no clear protocol of preoperative and postoperative testing, leading to inconsistent preoperative and postoperative evaluations. Intraoperative findings were based on operative notes and presence of symptoms was based on clinic notes alone, again reflecting the retrospective nature of the study. There was also lack of complete follow-up in some subjects.

CONCLUSIONS

In conclusion, focused TTE can be diagnostic in accurately identifying intramural AAOCA in pediatric patients. Unroofing can be performed safely with minimal early morbidity, but symptoms can persist, including rare life-threatening events, without evidence of reversible ischemia by postoperative provocative testing. This study highlights that, although surgery can be safely performed on children in whom the preoperative risk of myocardial ischemia is frequently unknown, risk stratification for subsequent events after unroofing is also difficult to assess in the absence of standardized protocols.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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