Anomalous Aortic Origin of a Coronary Artery

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ABSTRACT: Anomalous aortic origin of a coronary artery (AAOCA) is the second leading cause of sudden cardiac death in young athletes. The pathophysiology leading to sudden cardiac death, the specific risks associated with the different varieties of AAOCA, and the effects of different management strategies on the risk of sudden cardiac death are all unknown. This article describes the current knowledge of AAOCA, a proposed nomenclature for the different anatomic subtypes, the different modalities used to diagnose and characterize the disease, the available management strategies, and an algorithm used by the authors to diagnose and manage these patients.

INTRODUCTION

Congenital or acquired coronary artery anomalies are not infrequently diagnosed in children and adolescents, many of whom actively participate in routine exercise and/or competitive sports. These anomalies include anomalous aortic origin of a coronary artery (AAOCA), myocardial bridge or intramyocardial coronary artery, anomalous origin of the right or left coronary artery from the pulmonary artery, and sequelae of Kawasaki disease. This review focuses on AAOCA, the second leading cause of sudden cardiac death (SCD) in young US athletes[1] and a condition that poses challenges to risk stratification and optimal patient management.

AAOCA is a congenital abnormality of the origin or course of a coronary artery that arises from the aorta. This condition is associated with SCD, especially when the anomalous coronary originates from the opposite sinus of Valsalva. The clinical manifestations of patients presenting with AAOCA are quite variable, ranging from evident myocardial ischemia, such as angina-like chest pain and sudden cardiac arrest (SCA), to complete lack of symptoms. The exact mechanisms leading to SCA or SCD and the absolute determinants of risk are not completely understood. Risk stratification is frequently determined through myocardial functional studies to assess for evidence of inducible ischemia during provocative testing. Some patients may require invasive assessment with cardiac catheterization, including intravascular ultrasound (IVUS) and measurement of fractional flow reserve (FFR).

For the past 5 to 10 years, many efforts have advanced the knowledge about AAOCA, including specific anatomic types, diagnostic evaluation with imaging and myocardial functional assessment, and management strategies.[2,3] For the last several years, speakers from leading institutions have held dedicated scientific forums to discuss coronary anomalies. It is believed that prospective data gathering, longitudinal follow-up, and accrued experience will lead to better understanding of this condition and will directly benefit counseling and shared decision making with patients and families affected by AAOCA. This review highlights the current knowledge around AAOCA and serves as a general guide for clinicians.

SUDDEN CARDIAC DEATH

Sudden cardiac death leads to substantial anxiety in schools, sports organizations, and communities in general, especially because most events occur unexpectedly in healthy children or young athletes during or immediately after exercise.[1,4-7] Quite often, SCD is the first symptom of underlying cardiac disease. The risk of SCD in athletes is estimated at between 0.5 and 1 per 100,000 athlete-years,[8] though more recent studies have reported a higher incidence in specific populations.[9] However, the risk for all young individuals may be much higher. For example, the Resuscitation Outcomes Consortium reported an incidence of 8 cardiac arrests per 100,000 person-years in individuals under 21 years of age.[10]

Data from National Collegiate Athletic Association athletes have shown that SCD accounts for 16% of deaths compared to accidents (51%), suicides (9%), and homicides (6%).[9] The most common cause of SCD was notably unknown in 31% of these athletes followed by anomalous coronary arteries in 14%; interestingly, hypertrophic cardiomyopathy (HCM), dilated nonischemic cardiomyopathy, and myocarditis each accounted for 8%. By contrast, data from the US National Registry of Sudden Death in Athletes shows that the most frequent related causes were HCM (36%), congenital coronary anomalies (19%), indeterminate with left ventricular hypertrophy (9%), and myocarditis (7%).[11] In military recruits, the SCD incidence appears higher (13 per 100,000 person-years), with anomalous coronary arteries reported in 33%, myocarditis in 20%, and

HCM in 13%.[12] Males appear to be more likely to suffer SCD than females,[13] and nonwhites are overrepresented in SCD registries compared to whites.[1] In fact, most recent studies have suggested a higher occurrence of SCD in the black race, with no known reason.[9,11,13]

Unquestionably, congenital coronary anomalies are among the most frequent causes of SCD in youth. An increasing number of anomalies are being incidentally found on imaging studies that are done for preparticipation evaluations, for other common reasons (such as presence of a murmur or an "abnormal" electrocardiogram), or as part of screening programs to detect cardiovascular conditions possibly associated with SCD, as is currently ongoing in Texas with cardiac magnetic resonance (CMR) imaging.[14,15] About 75% of SCDs are related to a cardiovascular etiology. They usually occur during or after exercise and are most commonly associated with dynamic exercise.[1]

The pathophysiological mechanisms that predispose individuals to SCD, the risk conferred by different anatomical subtypes, and the effect of current treatment strategies on reducing SCD risk are not fully known. Several pathophysiological mechanisms have been postulated to lead to SCA or SCD in those with AAOCA, including the presence of coronary ostial abnormalities, compression of an interarterial segment between the great vessels, compression of an intramural segment during exercise, and obstruction by a flap-like ridge related to an acutely angulated coronary artery—all leading to myocardial ischemia and development of ventricular arrhythmia.[16-18] However, it is not fully understood what ultimately leads to a sudden cardiac event given that these patients often performed similar levels of exercise for many years without symptoms.

ANATOMY AND NOMENCLATURE

AAOCA is the abnormal origin of one or more coronary arteries from the aorta, and it encompasses a wide spectrum of anatomical variations. Despite an increased understanding about this pathology, it is unclear which pathophysiological mechanisms are related to SCD and which types of AAOCA confer the highest risk. Due to the wide anatomical variability, a systematic description of the anatomy is necessary to get a better understanding of AAOCA.[19]

Ostial Location

The topographic location of the ostium can be described using the circumferential location and height of the ostium. Figure 1 depicts a proposed system to describe the precise location of each coronary ostium with respect to the sinuses and commissures of the aorta. Based on this nomenclature system, the normal right coronary artery arises from a 1b-I location

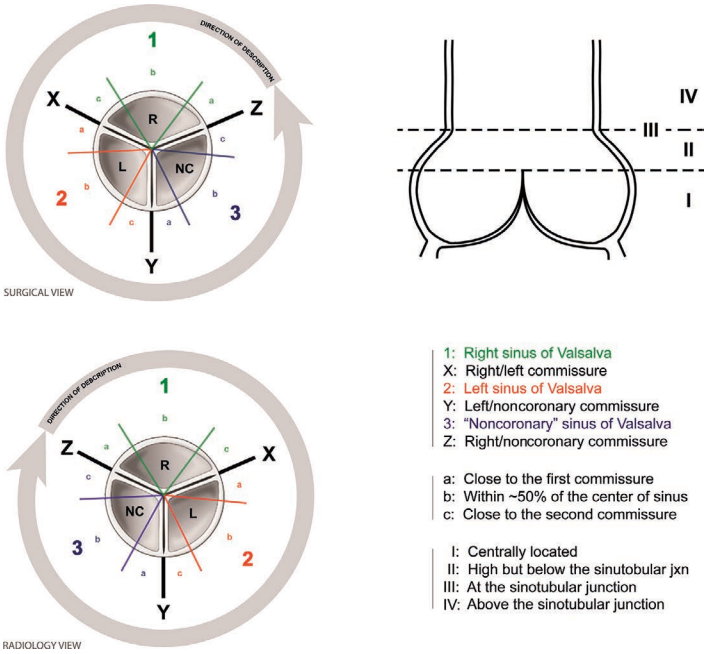

Figure 1.
Topography map to identify the location of the coronary ostia. Each sinus is indicated by a number, and the radial location of the ostium within the sinus is indicated by a letter. The height of the ostium in the aortic root/ascending aorta is indicated by a Roman numeral. The normal location of the left main coronary ostium is 2b-I and the normal location of the right coronary ostium is 1b-I. © 2013 Texas Children's Hospital (reprinted with permission).

whereas a normal left coronary artery arises from a 2b-I location. In our experience, the vast majority of AAOCA variants appear to arise high and on the opposite side of the intercoronary commissure (ie, location 2a-III/IV for an anomalous right coronary and location 1c-III/IV for an anomalous left coronary). Additional coronaries can be described in a similar fashion, such as an anomalous circumflex arising directly from the aorta.

Ostial Relationship

The spatial relationship between two coronary ostia is important as it may determine whether both coronary vessels represent branches from a single coronary or two completely separate vessels. The ostial relationship can be graded from grade 1 (separate coronary arteries) to grade 4 (single coronary artery with separate branches) (Figure 2).

Ostial Morphology

Almost all anomalous coronaries have to take a more angled course proximally as the vessel travels to its destination. In

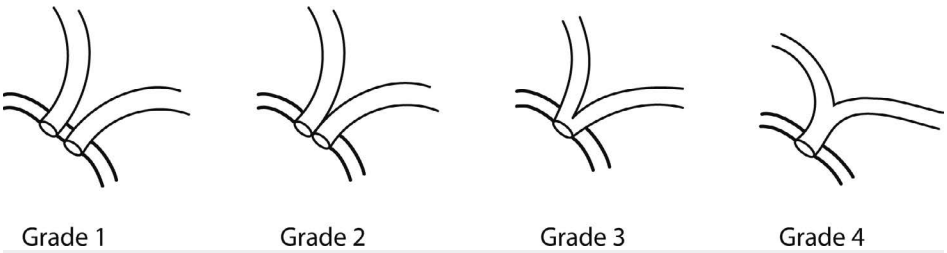

Figure 2.
Nomenclature for the relationship between two coronary ostia. Grade 1: two separate ostia; grade 2: separate but adjacent ostia; grade 3: common ostium with bifurcation within the aortic wall; grade 4: single coronary with bifurcation outside of the aortic wall. © 2018 The University of Texas Dell Medical School (reprinted with permission).

simple terms, a coronary ostium can be described as round, slit-like (if the anteroposterior dimension is shorter than the superoinferior dimension), and/or stenotic (if the ostium is smaller than the distal coronary).

Course

The interarterial segment of a coronary artery travels between the aorta and the pulmonary artery. An intramural segment travels within the wall of the aorta before arising from it. An intramyocardial coronary segment is said to be present when the vessel is completely surrounded by myocardium. Myocardial bridges, where the coronary is surrounded by myocardium after having had an initial epicardial course, are a relatively common type of intramyocardial coronary. An unusual variant of an intramyocardial vessel is the intraseptal anomalous left coronary artery. This is usually characterized by a single coronary artery arising from the right sinus (or an anomalous left coronary artery arising from the right sinus), with the left coronary diving into the interventricular septum below the level of the pulmonary valve (and behind the right ventricular outflow tract). The anomalous vessel will usually resurface lateral to the pulmonary artery before bifurcating into the anterior descending and circumflex branches.

The Intercoronary Pillar

An underappreciated anatomical feature of the aortocoronary complex is the intercoronary pillar, which is a thickening of tissue that extends cranially from the intercoronary commissure up to the sinotubular junction; it can be quite thick in some patients (Figure 3). This pillar likely contributes to the support of the aortic valve. Furthermore, we believe that this thick structure may play a significant role in compressing the anomalous coronary traveling behind it. As such, any surgical intervention performed to treat AAOCA should aim to alter the anatomy such that the ostium is appropriately away from the intercoronary pillar.

DIAGNOSIS AND EVALUATION

An increasing number of children and adolescents are being diagnosed with AAOCA following routine pre-participation screening, detection of a murmur, or an abnormal electrocardiogram (ECG) rather than during evaluation of symptoms. Evaluation prompts additional imaging, typically an echocardiogram, and the suggested diagnosis of AAOCA is then confirmed by advanced imaging, ie, computerized tomoraphy angiography (CTA) or CMR.

Patients with AAOCA may present with variable degrees of symptoms, although half are asymptomatic upon presentation. Concerning symptoms include chest pain (especially on exertion), dyspnea, palpitations, and syncope (especially on exertion).[1,5,11,13,18] In a study by Basso et al. of 27 individuals who experienced SCD due to AAOCA, only 10 presented with symptoms prior to the event.[5] An acute angle of take-off and slit-like ostium was evident in all 10 cases. In a series published by Eckart et al., 52% of military recruits who experienced SCD reported previous symptoms of chest pain, dyspnea, and syncope.[12] In our experience with AAOCA, 51% of patients were asymptomatic, 29% had chest pain, 15% had syncope, and 3.3% presented with SCA.[20] Of those presenting with SCA, one had a previous diagnosis of asthma and used his inhaler when he felt short of breath during exercise, one had been evaluated for syncope following exertion that was deemed vasovagal in nature, and two were completely asymptomatic.

Advanced Imaging

Cross-sectional imaging by CTA or CMR is necessary to confirm the diagnosis and, more importantly, to accurately define the anatomy of the anomalous vessel, including interarterial and/or intramural course, intraseptal or intramyocardial course, and ostial morphology.

Coronary CTA provides precise spatial resolution with excellent definition of ostial morphology and location of the anomalous coronary artery's proximal course (Figure 4). Moreover, it does not require sedation in younger children, and the amount of ionizing radiation has significantly decreased with new-generation scanners. The protocol includes retrospective ECG-gated dynamic CTA of the heart, and images are post-processed using a 3-dimensional (3D) workstation, with virtual angioscopy providing great details of ostial morphology. The presence and

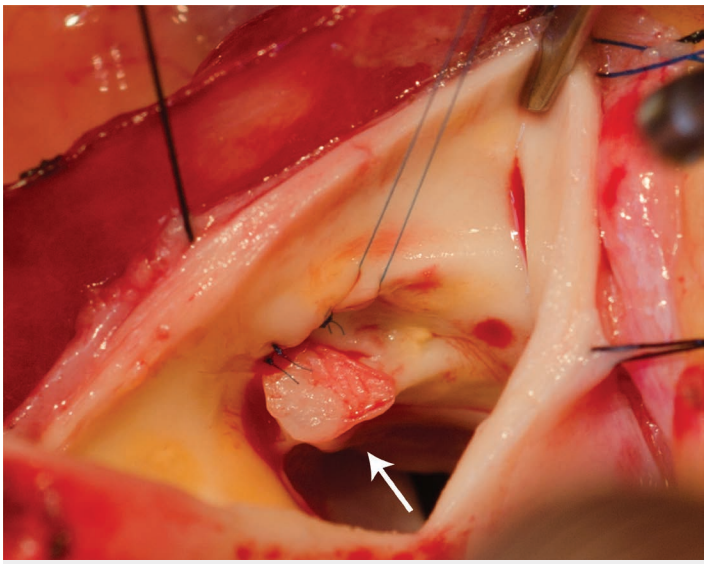

Figure 3.
Intraoperative image of a patient with an anomalous left coronary artery and a very thick intercoronary pillar (arrow). The intercoronary pillar likely plays an important role in the compression of the anomalous coronary artery that travels behind. © 2015 Texas Children's Hospital (reprinted with permission).

length of intramurality is obtained by analyzing the morphology of the proximal coronary artery, including the diameter of the vessel as it is laterally compressed in its intramural segment and the presence or lack of pericoronary mediastinal fat.[21] Coronary CTA is our preferred modality for advanced imaging after an initial diagnosis by echocardiography.

Some institutions prefer CMR imaging because it does not use ionizing radiation and provides accurate data on myocardial perfusion and viability, function, and flow patterns. However, this modality often requires sedation in younger children and lacks the spatial resolution needed to determine ostial morphology and course of the anomalous vessels.

Myocardial Functional Assessment

Exercise stress test (EST) is universally used to evaluate these patients, but there is controversial data regarding its validity. Studies have reported that 6% to 22% of patients who undergo surgery or experience SCD present with an abnormal EST.[5,22,23] In our program, 8% of patients have an abnormal EST.[23]

Imaging with nuclear perfusion stress (NPS) is typically used to evaluate myocardial perfusion; however, this technique is marred by false positive and false negative results, thus affecting its reliability to truly identify myocardial ischemia. Abnormal perfusion defects with this technique have been reported in patients with AAOCA, including following surgical intervention.[22] We have observed similarly poor reliability with NPS compared to stress CMR, with sensitivity of only 33%.[24]

Stress CMR imaging with pharmacologic agents has demonstrated superiority to NPS in assessing myocardial perfusion in patients with AAOCA.[24] This technique appears to be feasible, safe, and well tolerated in the pediatric population.[24] It also reliably detects perfusion defects and wall motion abnormalities, which contributes to risk stratification in these patients.

Cardiac Catheterization

Although cardiac catheterization is primarily used in the adult population, it recently has been shown to be feasible and safe in pediatric patients with AAOCA.[25] Angelini et al. proposed an approach to risk-stratify the more common anomalous right coronary artery arising from the opposite sinus of Valsalva, in which the degree of stenosis in the intramural segment is determined by the use of IVUS throughout the cardiac cycle.[26] We have used cardiac catheterization with IVUS and FFR measurement with adenosine and/or dobutamine for risk stratification, and it has allowed us to identify significant decrease in the lumen and flow of coronary arteries with an intramyocardial segment in the setting of both AAOCA and myocardial bridges.[25] Our initial experience has been published elsewhere.[25] We have mainly used cardiac catheterization with IVUS/FFR in patients with long intramyocardial or intraseptal course of the coronaries and concerning symptoms or signs of ischemia and in patients with AAOCA and unclear anatomy. Due to the absence of guidelines regarding normal values in pediatric patients, we have used parameters similar to the ones used in adult patients with coronary artery disease (FFR < 0.8 after administration of adenosine and/or dobutamine). The validity of this threshold remains to be validated in AAOCA patients, but we have observed patients with abnormal FFR prior to surgical intervention return to normal values following surgery (unpublished data). Additionally, we have looked at the correlation between FFR and stress CMR and demonstrated good correlation between abnormal FFR measurements and perfusion abnormalities on stress CMR (manuscript in preparation).

CLINICAL DECISION MAKING AND MANAGEMENT

The optimal management strategy for patients with AAOCA remains controversial given the many uncertainties related to risk factors according to anatomic subtypes, the true risk for SCD over a lifetime, and the longitudinal effect of intervention versus nonintervention. Based on the many unknowns and the

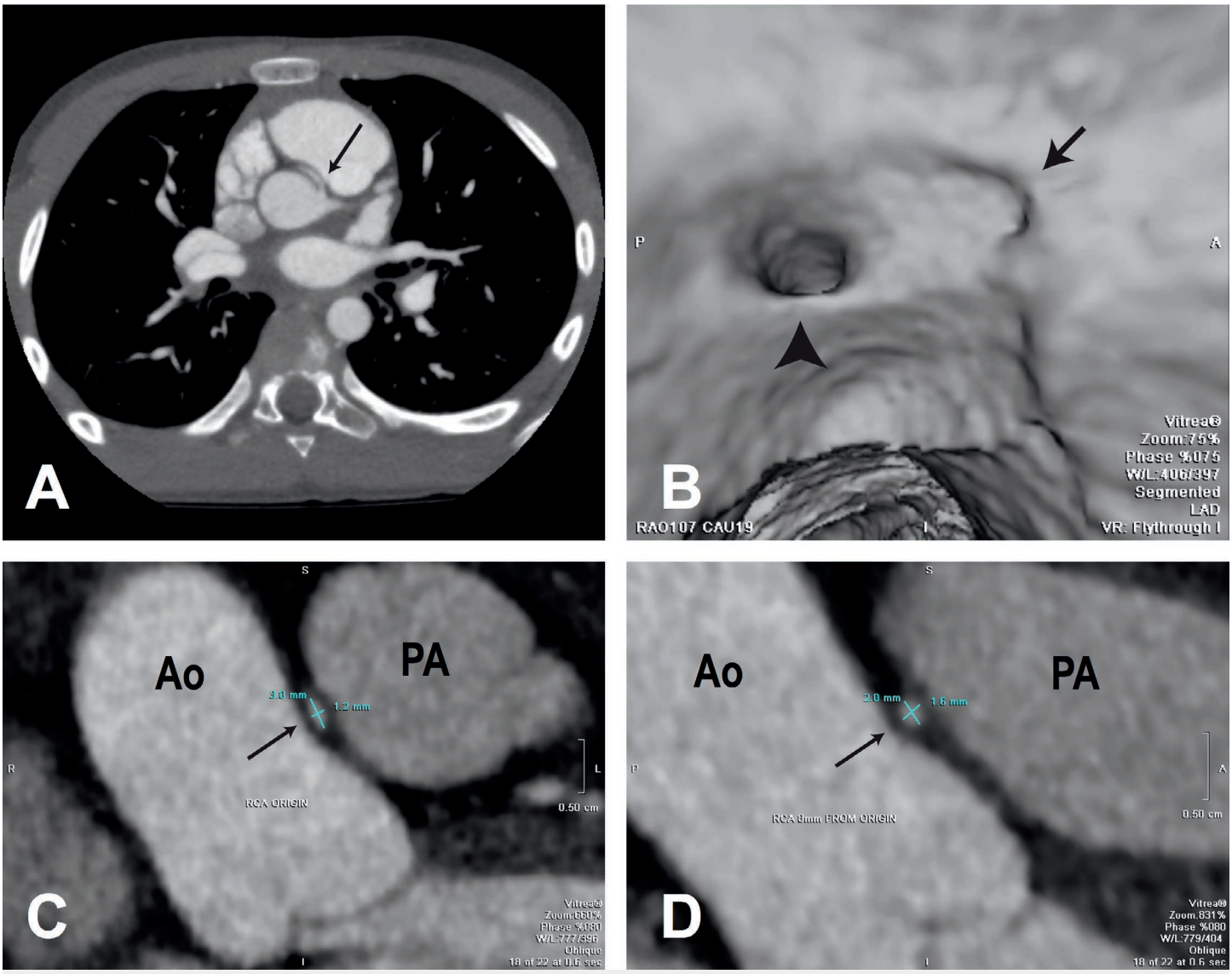

Figure 4.
Computerized tomographic angiography demonstrating an anomalous right coronary artery. (A) The anomalous right coronary arises from the left sinus and travels intramurally and in between the aorta and the pulmonary artery. (B) A virtual angioscopy shows a normal left coronary ostium (arrowhead) and the anomalous right coronary with a stenotic slit-like ostium arising just above and to the left of the intercoronary commissure. (C) The anomalous coronary (arrow) has an oval shape on its intramural segment compared to (D) the round shape of the distal coronary past its intramural segment. © 2014 Texas Children's Hospital (reprinted with permission). Ao: aorta; PA: pulmonary artery

need for meaningful data, efforts have been made nationwide to develop programs to evaluate and manage these patients.[2,27] Our team has developed a dedicated Coronary Anomalies Program (CAP) to evaluate and manage these patients more consistently using a standardized approach to diagnosis, management, and follow-up. A clinical algorithm for workup and management was developed based on the available data and consensus of a multidisciplinary team of pediatric and adult cardiologists, interventional cardiologists, surgeons, cardiovascular radiologists, cardiovascular anesthesiologists, nurses, and research and outcomes staff (Figure 5).

As part of the algorithm, patients with AAOCA who are referred to the CAP are evaluated by a core group of pediatric cardiologists and undergo standardized testing including electrocardiography, echocardiography, advanced imaging

Figure 5.
Algorithm for diagnosis and management of anomalous aortic origin of a coronary artery at Texas Children's Hospital. © 2018 Texas Children's Hospital (reprinted with permission). ALCA-R: anomalous left coronary from the right sinus; ALCx: anomalous left circumflex artery; ARCA-L: anomalous right coronary from the left sinus: CAP: Coronary Anomalies Program; CTA: computerized tomographic angiography; MRI: magnetic resonance imaging; IVUS: intravascular ultrasound; FFR: fractional flow reserve

[a] Consent obtained for participation in prospective Congenital Heart Surgeons' Society and Texas Children's Hospital databases.

[b] Additional studies may be performed depending on the clinical assessment.

[c] External echocardiograms do not need to be repeated if the study is deemed appropriate.

[d] CPET or stress cMRI are not necessary on patients who present with aborted sudden cardiac death. These studies may be deferred in young patients.

[e] An external CTA may be used if images can be uploaded and the study provides all necessary information to make a decision. These studies may be deferred in patients < 8 years unless there are clinical concerns.

[f] An intraseptal coronary is an abnormal vessel (usually a left coronary arising from the right sinus) that travels posteriorly into the septum below the level of the pulmonary valve.

[g] Unroofing if significant intramural segment, neo-ostium creation, or coronary translocation if intramural segment is behind a commissure, coronary translocation if short or no intramural segment. Surgical intervention will be offered for these patients between ages 10-35; other patients will be considered on a case-by-case basis. Aspirin will be administered for 3 months after surgery.

[h] Restriction from participation in all competitive sports and in exercise with moderate or high dynamic component (eg, soccer, swimming, tennis, basketball, football).

[i] Patient may be seen by outside primary cardiologist.

[j] Postoperative patients will be cleared for exercise and competitive sports based on findings at the third month postoperative visit, including results of CPET, stress cMRI, and CTA.

with retrospectively ECG-gated CTA, and myocardial functional studies, specifically EST and stress CMR. In a small number of patients, cardiac catheterization with IVUS and FFR is also indicated. The CAP team holds biweekly multidisciplinary meetings to review and discuss patients' data and decide on the best management. In general, patients with an anomalous left coronary artery and high-risk anatomy (origin from the opposite sinus with interarterial +/- intramural course) are offered surgical intervention. Patients with other coronary anomalies are offered surgical intervention if they have concerning characteristics such as symptoms clearly ascribed to ischemia, a positive functional test, or high-risk anatomy such as a long intramural course and ostial abnormalities, regardless of symptomatology. Patients undergoing surgical intervention are placed on low-dose aspirin on postoperative day 1 and continued for 3 months postoperatively. Troponin levels are not routinely trended in these patients.

In general, given the lower perceived risk of SCA in very young patients, we tend to defer surgical intervention in pediatric patients until they are older than 10 years of age unless they have evidence of ischemia. The threshold for surgical intervention is similarly higher for asymptomatic patients older than 35 years of age since they also seem to have a lower risk of SCA.

All patients are followed at particular time intervals. Patients who undergo surgical intervention are re-evaluated after 3 months with electrocardiography, functional testing, and CTA. If the workup is reassuring, patients are allowed to return to full exercise activities. In general, exercise restriction is only recommended for patients who are awaiting surgical intervention, who are currently in the postoperative period, who have high-risk lesions and refuse surgical intervention, or who have ischemic symptoms or positive functional testing but whose anatomy is unsuitable for surgical intervention. Patients with low-risk lesions that do not warrant surgical intervention are not exercise restricted.

Since the majority of SCD events occur during or immediately after intense exercise,[1] exercise restriction has been recommended as a management strategy to prevent SCD in some patients with AAOCA.[28] However, the effectiveness of this strategy is unclear due to reports of patients suffering from SCD while engaged in minimal activity.[29] In addition, exercise restriction is not a benign strategy since it increases long-term cardiovascular risks and likely has a significantly negative psychosocial impact in young athletes.[30] Based on a decision analysis by our group, we found that exercise restriction was not a preferred long-term strategy in any cohort of patients with AAOCA due to its detrimental long-term effect.[31] We only recommend exercise restriction in patients awaiting surgical intervention and those who have clinical symptoms or ischemic findings during exercise.

SURGICAL TECHNIQUES AND OUTCOMES

Multiple surgical procedures have been performed in an attempt to treat ischemic symptoms or prevent SCD in patients with AAOCA. However, without a clear understanding of the pathophysiological mechanisms of SCD in AAOCA, it is difficult to define the optimal surgical strategy for a particular patient.

Unroofing Procedure

When an intramural coronary segment is present, we use the unroofing procedure (Figure 6), which removes the intramural segment by excising the intervening wall between the lumens of the aortic and anomalous coronary. The procedure also increases the size of the ostium and essentially moves it to the correct sinus (Figure 7 A). The longer the intramural segment, the more effective the procedure is at relocating the ostium. However, if the intramural segment is relatively short (< 5 mm, in our experience), the procedure may fail to relocate the ostium to the correct sinus and/or the anomalous vessel may be at risk of compression by the intercoronary pillar (Figure 7 B). Therefore, a short intramural segment may require a different procedure. Although some authors have used the unroofing procedure with commissure takedown and resuspension when the intramural segment travels below the level of the aortic valve, we advise against it because of the long-term risk of developing aortic valve incompetence.[32]

Coronary Translocation

Coronary translocation entails dividing the coronary artery as it arises from the aortic wall and reimplanting it into the correct sinus. Some groups have advocated the universal use of this procedure because it closely resembles the normal coronary arrangement.[33] However, the procedure is technically more demanding, and the long-term results of creating a circumferential anastomosis of the coronary artery on the aorta are unknown. As such, we reserve this procedure for cases in which the intramural segment travels below the level of the aortic valve or for patients with an absent or short intramural segment (Figure 7).

Creation of Neo-Ostium

This procedure is used for patients with a long intramural segment that travels below the level of the aortic valve. It entails unroofing the portion of the intramural segment that sits within the correct sinus while leaving the rest of the intramural

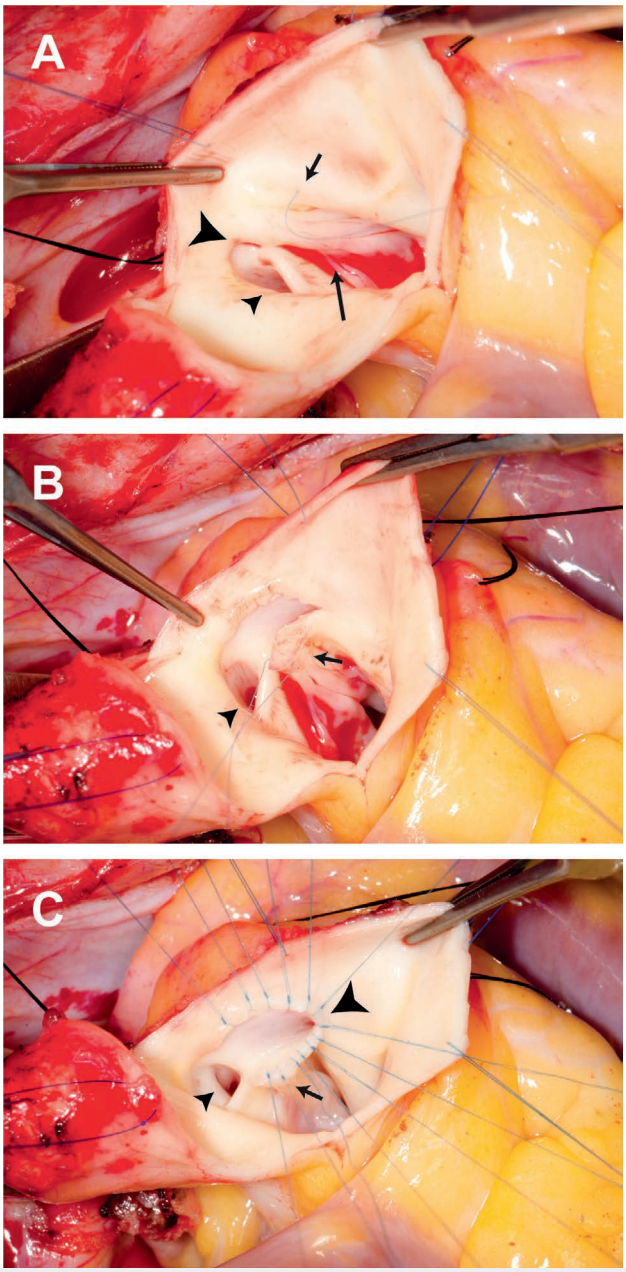

Figure 6.
Unroofing procedure for an anomalous right coronary artery from the left sinus. (A) The anatomy prior to unroofing. The left coronary ostium (small arrowhead) is large, round, and normal (2b-I). The right coronary ostium (large arrowhead) is stenotic and slit-like, just above and rightward to the intercoronary commissure (long arrow), located at 2a-II. The fine suture (small arrow) is placed transmurally and indicates the site where the right coronary arises out of the aorta externally. (B) The intramural segment has been unroofed. A medium-thickness intercoronary pillar (small arrow) can be seen. (C) The end results after placement of tacking sutures around the unroofed segment. The right coronary ostium (large arrowhead) is now wide open and away from the intercoronary pillar (small arrow). © 2018 Texas Children's Hospital (reprinted with permission).

segment (behind the aortic valve) alone.[34] The intramural segment within the correct sinus has to be long enough to allow for an adequately sized neo-ostium.

Pulmonary Translocation

Based on the premise that the presence of an interarterial segment between the aorta and the pulmonary artery may be a pathophysiological mechanism of SCD, some groups have advocated the use of a lateral or anterior pulmonary translocation for patients with a single coronary artery or those with no intramural segment.[35,36] These procedures increase the available space between the aorta and the pulmonary artery.[37]

Other Procedures

Several other procedures have been described to treat AAOCA, including coronary artery plasty,[38] unflooring/unroofing procedure (ie, performing both an unroofing and a patch plasty),[39] and coronary artery bypass grafting, although this is likely an ineffective procedure for most patients with AAOCA because of the very intermittent nature of ischemia and the consequent risk of thrombosis due to competitive flow. Other groups have suggested using ostial coronary artery stenting in the catheterization lab to prevent compression of the proximal segment of the anomalous coronary in adults.[26] The long-term results of such a strategy, especially in young patients, is unknown.

Surgical Results

Multiple retrospective and prospective surgical series have shown that surgical intervention for AAOCA can be performed with relatively low perioperative risk and good mid-term outcomes.[40,41] However, there have been reports of patients suffering from cardiac events following surgical intervention for AAOCA.[42,43] The cause of these events is unclear but may represent persistent compression of the coronary artery by a residual intramural segment or by the intercoronary pillar,[43] the existence of an arrhythmogenic myocardium due to chronic ischemia, or other poorly understood mechanisms that translate into persistent risk of SCD.

FUTURE DIRECTIONS

Many questions still remain regarding the diagnosis and management of patients with AAOCA: Who is at risk of SCD? What is the actual risk? What is the optimal strategy for a particular patient? Only by developing strategies to standardize workup and follow-up of patients with AAOCA will we be able to better understand this disease and come closer

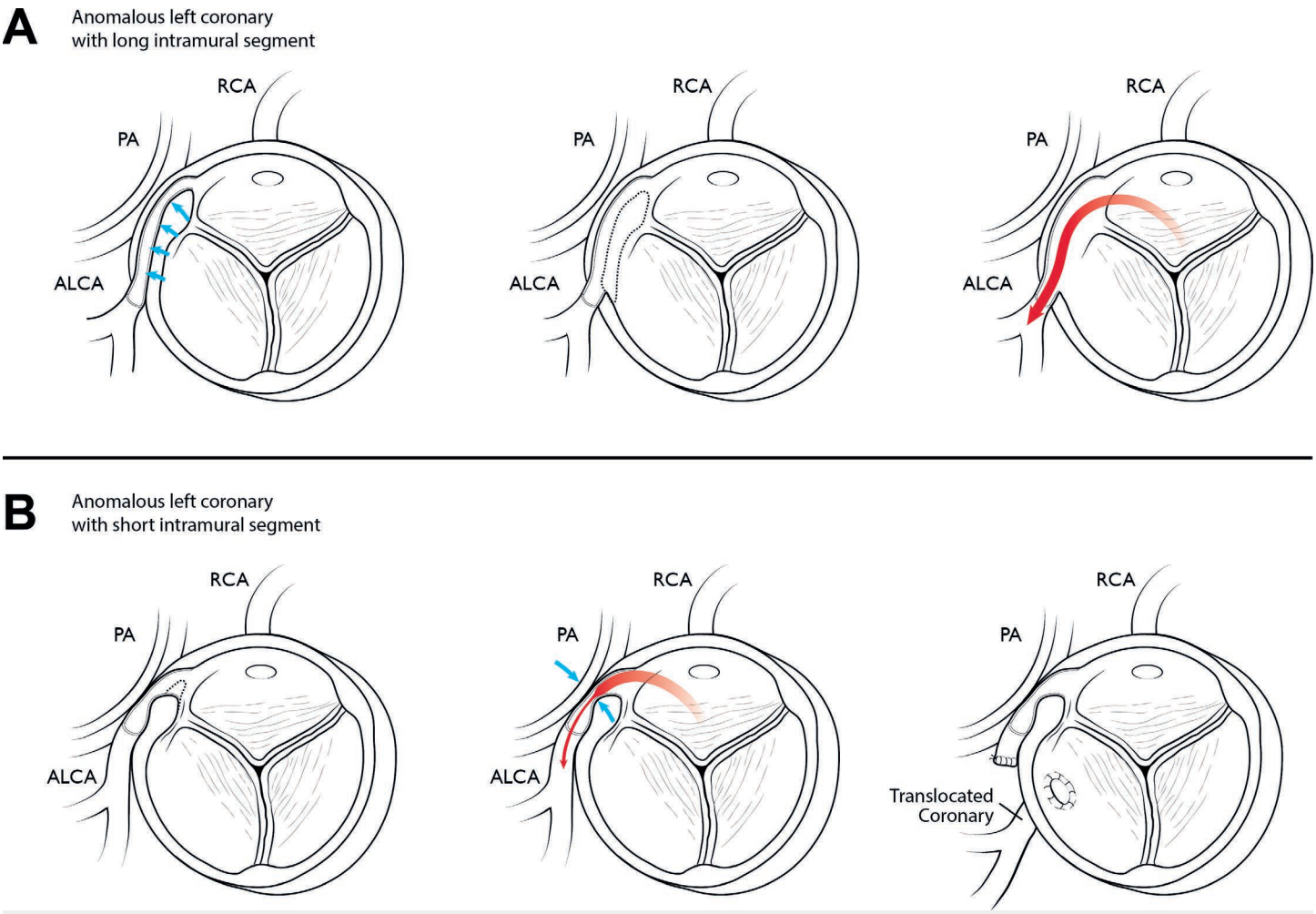

Figure 7.
Diagram illustrating the result of unroofing an anomalous coronary artery with a long and a short intramural segment. (A) If a there is a long intramural segment, unroofing eliminates the intramural segment, enlarges the ostium, and effectively moves the ostium to the correct sinus. (B) On the contrary, if it the intramural segment is short, unroofing eliminates the intramural segment but the ostium remains arising from the incorrect sinus and the coronary may still be compressed between the thick intercoronary pillar and the pulmonary artery. In this case, coronary translocation may be a better alternative than unroofing.
 RCA: right coronary artery; PA: pulmonary artery; ALCA: anomalous left coronary artery

to defining the optimal management strategy for individual patients. Initiatives such as the multi-institutional Congenital Heart Surgeons' Society AAOCA Registry,[2] dedicated programs in academic institutions,[3,27] and other multicenter collaborations will help advance our understanding of this disease.

In addition, all of the uncertainty can have a significant impact on the emotional and psychosocial wellbeing of patients and families with AAOCA, and this aspect has been poorly studied. It is incumbent upon us to develop protocols to study the psychosocial implications of this diagnosis and to design effective strategies to mitigate these concerns as we move the field forward.

Conflict of Interest Disclosure:
The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords:
anomalous aortic origin of a coronary artery, AAOCA, coronary anomaly, sudden cardiac death

KEY POINTS

- Optimal risk stratification for patients with anomalous aortic origin of a coronary artery (AAOCA) is still lacking, though substantial advancements have occurred in the past few years.
- Anomalous right coronary artery from the opposite (left) sinus of Valsalva with an intramural course is likely, but not always, benign in most cases.
- Surgical treatment for AAOCA should be individualized depending on the particular anatomic characteristics and should aim to address all possible anatomic culprits.
- Prospective data gathering, longitudinal follow-up, and collaboration among centers will continue to foster a deeper understanding of this condition and how to better counsel patients and families.

REFERENCES

1. Maron BJ, Doerer JJ, Haas TS, Tierney DM, Mueller FO. Sudden deaths in young competitive athletes: analysis of 1866 deaths in the United States, 1980-2006. Circulation. 2009 Mar 3;119(8):1085-92.

2. Brothers JA, Gaynor JW, Jacobs JP, et al. The Registry of Anomalous Aortic Origin of the Coronary Artery of the Congenital Heart Surgeons' Society. Cardiol Young. 2010 Dec;20 Suppl 3:50-8.

3. Molossi S, Agrawal H. Coronary artery anomalies: A multidisciplinary approach to shape the landscape of a challenging problem. Congenit Heart Dis. 2017 Sep;12(5):596.

4. Stecker EC, Reinier K, Marijon E, et al. Public health burden of sudden cardiac death in the United States. Circ Arrhythm Electrophysiol. 2014 Apr;7(2):212-7.

5. 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 May;35(6):1493-501.

6. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol. 1992 Sep;20(3):640-7.

7. Link MS, Estes NA. Athletes and arrhythmias. J Cardiovasc Electrophysiol. 2010 Oct;21(10):1184-9.

8. Maron BJ. Sudden death in young athletes. N Engl J Med. 2003 Sep 11;349(11):1064-75.

9. Harmon KG, Asif IM, Klossner D, Drezner JA. Incidence of sudden cardiac death in National Collegiate Athletic Association athletes. Circulation. 2011 Apr 19;123(15):1594-600.

10. Atkins DL, Everson-Stewart S, Sears GK, et al.; Resuscitation Outcomes Consortium Investigators. Epidemiology and outcomes from out-of-hospital cardiac arrest in children: the Resuscitation Outcomes Consortium Epistry-Cardiac Arrest. Circulation. 2009 Mar 24;119(11):1484-91.

11. Maron BJ, Haas TS, Ahluwalia A, Murphy CJ, Garberich RF. Demographics and Epidemiology of Sudden Deaths in Young Competitive Athletes: From the United States National Registry. Am J Med. 2016 Nov;129(11):1170-7.

12. Eckart RE, Scoville SL, Campbell CL, et al. Sudden death in young adults: a 25-year review of autopsies in military recruits. Ann Intern Med. 2004 Dec 7;141(11):829-34.

13. Maron BJ, Haas TS, Murphy CJ, Ahluwalia A, Rutten-Ramos S. Incidence and causes of sudden death in U.S. college athletes. J Am Coll Cardiol. 2014 Apr 29;63(16):1636-43.

14. Angelini P, Vidovich MI, Lawless CE, et al. Preventing sudden cardiac death in athletes: in search of evidence-based, cost-effective screening. Tex Heart Inst J. 2013;40(2):148-55.

15. Angelini P, Cheong BY, Lenge De Rosen VV, et al. High-Risk Cardiovascular Conditions in Sports-Related Sudden Death: Prevalence in 5,169 Schoolchildren Screened via Cardiac Magnetic Resonance. Tex Heart Inst J. 2018 Aug 1;45(4):205-13.

16. Brothers J, Carter C, McBride M, Spray T, Paridon S. Anomalous left coronary artery origin from the opposite sinus of Valsalva: evidence of intermittent ischemia. J Thorac Cardiovasc Surg. 2010 Aug;140(2):e27-9.

17. Cox ID, Bunce N, Fluck DS. Failed sudden cardiac death in a patient with an anomalous origin of the right coronary artery. Circulation. 2000 Sep 19;102(12):1461-2.

18. Cheitlin MD, MacGregor J. Congenital anomalies of coronary arteries: role in the pathogenesis of sudden cardiac death. Herz. 2009 Jun;34(4):268-79.

19. Agrawal H, Mery CM, Krishnamurthy R, Molossi S. Anatomic types of anomalous aortic origin of a coronary artery: A pictorial summary. Congenit Heart Dis. 2017 Sep;12(5):603-6.

20. Molossi S, Mery C, Krishnamurthy R, et al. Standardized Approach to Patients with Anomalous Aortic Origin of a Coronary Artery: Results from the Coronary Anomalies Program at Texas Children's Hospital. J Am Coll Cardiol. 2015 Mar;65(10 Suppl):A501.

21. Krishnamurthy R, Masand P, Jadhav S, et al. Diagnostic Accuracy of Ct Angiography (CTA) for Critical Pathologic Features in Anomalous Aor-

tic Origin of the Coronary Arteries (AAOCA) in Children: A Comparative Study with Surgery in a Single Center. J Am Coll Cardiol. 2015 Mar 17;65(10 Suppl):A1304.

22. Brothers JA, McBride MG, Seliem MA, et al. Evaluation of myocardial ischemia after surgical repair of anomalous aortic origin of a coronary artery in a series of pediatric patients. J Am Coll Cardiol. 2007 Nov 20;50(21):2078-82.

23. Agrawal H, Mery C, Krishnamurthy R, et al. Stress Myocardial Perfusion Imaging in Anomalous Aortic Origin of a Coronary Artery: Results Following a Standardized Approach. J Am Coll Cardiol. 2017 Mar 1;69(11 Suppl):1616.

24. Noel C, Molossi S, Krishnamurthy R, Moffett B, Krishnamurthy R. Cardiac MR Stress Perfusion with Regadenoson and Dobutamine in Children: Single Center Experience in Repaired and Unrepaired Congenital and Acquired Heart Disease. Circulation. 2016 Apr;67(13 Suppl):964.

25. Agrawal H, Molossi S, Alam M, et al. Anomalous Coronary Arteries and Myocardial Bridges: Risk Stratification in Children Using Novel Cardiac Catheterization Techniques. Pediatr Cardiol. 2017 Mar;38(3):624-30.

26. Angelini P, Uribe C, Monge J, Tobis JM, Elayda MA, Willerson JT. Origin of the right coronary artery from the opposite sinus of Valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. Catheter Cardiovasc Interv. 2015 Aug;86(2):199-208.

27. Mery CM, Lawrence SM, Krishnamurthy R, et al. Anomalous aortic origin of a coronary artery: toward a standardized approach. Semin Thorac Cardiovasc Surg. 2014 Summer;26(2):110-22.

28. Peñalver JM, Mosca RS, Weitz D, Phoon CK. Anomalous aortic origin of coronary arteries from the opposite sinus: a critical appraisal of risk. BMC Cardiovasc Disord. 2012 Oct 1;12:83.

29. Jo Y, Uranaka Y, Iwaki H, Matsumoto J, Koura T, Negishi K. Sudden cardiac arrest: associated with anomalous origin of the right coronary artery from the left main coronary artery. Tex Heart Inst J. 2011;38(5):539-43.

30. Wen CP, Wai JP, Tsai MK, et al. Minimum amount of physical activity for reduced mortality and extended life expectancy: a prospective cohort study. Lancet. 2011 Oct 1;378(9798):1244-53.

31. Mery CM, Lopez KN, Molossi S, et al. Decision analysis to define the optimal management of athletes with anomalous aortic origin of a coronary artery. J Thorac Cardiovasc Surg. 2016 Nov;152(5):1366-1375.e7.

32. Romp RL, Herlong JR, Landolfo CK, et al. Outcome of unroofing procedure for repair of anomalous aortic origin of left or right coronary artery. Ann Thorac Surg. 2003 Aug;76(2):589-95; discussion 595-6.

33. Law T, Dunne B, Stamp N, Ho KM, Andrews D. Surgical Results and Outcomes After Reimplantation for the Management of Anomalous Aortic Origin of the Right Coronary Artery. Ann Thorac Surg. 2016 Jul;102(1):192-8.

34. Karamichalis JM, Vricella LA, Murphy DJ, Reitz BA. Simplified technique for correction of anomalous origin of left coronary artery from the anterior aortic sinus. Ann Thorac Surg. 2003 Jul;76(1):266-7.

35. Rodefeld MD, Culbertson CB, Rosenfeld HM, Hanley FL, Thompson LD. Pulmonary artery translocation: a surgical option for complex anomalous coronary artery anatomy. Ann Thorac Surg. 2001 Dec;72(6):2150-2.

36. Mainwaring RD, Reddy VM, Reinhartz O, et al. Anomalous aortic origin of a coronary artery: medium-term results after surgical repair in 50 patients. Ann Thorac Surg. 2011 Aug;92(2):691-7.

37. Guerra VC, Recto MR, Goldman C, Yeh T Jr. Anomalous aortic origin of the coronary artery: does pulmonary artery translocation affect coronary artery course? J Thorac Cardiovasc Surg. 2013 Dec;146(6):1549-51.

38. Karl TR, Provenzano SC, Nunn GR. Anomalous aortic origin of a coronary artery: a universally applicable surgical strategy. Cardiol Young. 2010 Dec;20 Suppl 3:44-9.

39. Dekel H, Hickey EJ, Wallen J, Caldarone CA. Repair of anomalous aortic origin of coronary arteries with combined unroofing and unflooring technique. J Thorac Cardiovasc Surg. 2015 Aug;150(2):422-4.

40. Cheezum MK, Liberthson RR, Shah NR, et al. Anomalous Aortic Origin of a Coronary Artery From the Inappropriate Sinus of Valsalva. J Am Coll Cardiol. 2017 Mar 28;69(12):1592-1608.

41. Mery CM, De León LE, Molossi S, et al. Outcomes of surgical intervention for anomalous aortic origin of a coronary artery: A large contemporary prospective cohort study. J Thorac Cardiovasc Surg. 2018 Jan;155(1):305-319.e4.

42. Sachdeva S, Frommelt MA, Mitchell ME, Tweddell JS, Frommelt PC. Surgical unroofing of intramural anomalous aortic origin of a coronary artery in pediatric patients: Single-center perspective. J Thorac Cardiovasc Surg. 2018 Apr;155(4):1760-8.

43. Agrawal H, Sexson-Tejtel SK, Qureshi AM, et al. Aborted Sudden Cardiac Death After Unroofing of Anomalous Left Coronary Artery. Ann Thorac Surg. 2017 Sep;104(3):e265-e267.