Received: 11 May 2017 | Accepted: 20 May 2017
DOI: 10.1111/chd.12497

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Introduction to anomalous aortic origin of a coronary artery

Julie A. Brothers, MD[1,2]

[1]Division of Cardiology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA

[2]Perelman School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania, USA

Correspondence
Julie Brothers, MD, Department of Pediatrics, The Children's Hospital of Philadelphia, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA 19104.
Email: brothersj@email.chop.edu

Abstract

Anomalous aortic origin of a coronary artery (AAOCA) occurs when both coronary arteries arise from the same aortic sinus from a single ostium or two separate ostia. While most coronary anomalies are benign, the two most common subtypes that predispose to sudden cardiac death in the young are interarterial anomalous right coronary artery and interarterial anomalous left coronary artery. Practitioners face many challenges with AAOCA. Diagnosing patients may be difficult because children and adolescents are often asymptomatic and first presentation may be sudden death or sudden cardiac arrest. Risk stratification is also challenging as determining which unique characteristics place the child at highest risk of sudden death has not been adequately delineated and ischemic testing may give false negative results. Last, there is significant variability in decision making regarding management of youth with AAOCA. Future research is needed to help determine the best way to identify at-risk children and which treatment is the safest and most efficacious.

KEYWORDS

anomalous coronary artery, congenital heart disease, congenital heart surgery, pediatrics, sudden death

Anomalous aortic origin of a coronary artery (AAOCA) occurs when both coronary arteries arise from the same aortic sinus from a single ostium or two separate ostia. On occasion, the aberrant vessel may arise above the inappropriate sinus or the commissure instead of from the sinus itself. When the left main coronary artery (LCA) arises from the right sinus of Valsalva, we refer to this as ALCA-R and when the right coronary artery (RCA) arises from the left aortic sinus, we refer to this as ARCA-L. The anomalous coronary can subsequently course in different ways. The ALCA-R or ARCA-L can track anterior to the right ventricular outflow tract (RVOT) (prepulmonic) or posterior to the aorta (posterior or retroaortic); these are generally benign lesions although rare cases of ischemia have been reported with posterior ALCA-R.[1] A third subtype, intraseptal (intraconal/intramyocardial) ALCA-R, occurs when the anomalous left coronary artery courses caudad to the pulmonary valve, through the conal septum. The anomalous vessel may arise as a common trunk of the RCA or separately from the right sinus of Valsalva in close proximity to the RCA ostium. This variant is generally benign as well. The final subtype, interarterial, is most commonly associated with myocardial ischemia and sudden cardiac death (SCD).[2] The interarterial vessel is commonly intramural, traveling in the wall of the aorta with the anomalous vessel ostium located juxtacommissural or just above the commissure.

Because of the potential for SCD, there has been considerable interest in trying to determine the mechanism of ischemia. Different theories have been set forth; ischemia is believed to be the result of a combination of several factors that lead to coronary occlusion or compression over time which ultimately leads to myocardial ischemia and ventricular tachycardia or fibrillation. One idea as to the cause of ischemia is the acute angle takeoff and intramural segment of the proximal anomalous coronary artery that may flatten as the aorta dilates with exercise.[2–4] In a recent review of all SCD over an 18-year-period in the United Kingdom, there were 17 cases of AAOCA (0.7%), with 10 ARCA-L and 7 ALCA-R. All subjects had an interarterial course, 8 of which were intramural and 7 had an elliptical orifice.[5] As well, the majority of cases of sudden death in a large autopsy series of AAOCA had an acute angle takeoff from the aorta, with the exception of those with a single ostium.[3] As the anomalous interarterial coronary artery often arises in the pericommisural area, there is also the potential for flow limitation through this relatively noncompliant region. In adults, intravascular ultrasound (IVUS) has offered some insight into the pathophysiology, demonstrating hypoplasia and lateral luminal compression

of the proximal, intramural course of the coronary artery by the aorta. The compression is worse in systole and greatest during exercise.[6]

Indeed, one of the reasons there is much interest in AAOCA is that it is one of the leading causes of SCD but practitioners face many challenges when dealing with these patients. The first challenge is with actually diagnosing patients with this coronary anomaly as the physical examination and ECG are almost always normal. Patients frequently are diagnosed in the absence of symptoms but are found serendipitously when an echocardiogram is performed for other reasons, such as a heart murmur or abnormal ECG. When symptoms are present, they are frequently chest pain, palpitations, dizziness, and/or syncope at rest or during or with exercise. Unfortunately, for some, the first presentation is SCD or sudden cardiac arrest. Once the diagnosis is suspected, many young patients subsequently undergo a cardiac MRI or computed tomography angiography to better visualize the origin and proximal course of the coronary arteries. The modality chosen is largely provider- and institution-dependent, usually based on access to each imaging modality.

Once the diagnosis is made, we are currently unable to adequately risk stratify patients, especially those found serendipitously. Ideally, when a young person is diagnosed with AAOCA, we would be able categorize the patient to a high- or low-risk group. The high-risk group would be referred for surgery and/or exercise restriction and the low-risk group could potentially be cleared for competitive athletics. What do we need to do this? To a certain extent, we currently stratify patients by coronary artery course, in that most physicians do not recommend surgery or even exercise restriction in any coronary anomaly that does not course between the two great vessels. In those with an interarterial course, we would ideally be able to use features that we believe may place the patient at higher ischemic risk, such as a slit-like orifice, length of intramural course, vessel spasm, and intussusception. However, finding a unifying characteristic in sudden death cases has been challenging.[7] Angelini has used IVUS in adults to show that certain factors correlated with clinical severity: amount of hypoplasia of the intramural segment, amount of lateral compression of the intussusceptied segment, and lateral compression during exercise.[6] Unfortunately, the use of IVUS in children is not practical due to its invasive nature, lack of small enough catheter size, and lack of experience with this procedure in the pediatric population. A noninvasive way to evaluate for risk is needed in the pediatric population.

Another test commonly used for risk stratification is a maximal cardiopulmonary exercise test (CPET). The CPET is part of the evaluation after patients have been diagnosed with AAOCA and also for evaluation after surgery prior to clearing the patient for competitive athletics. However, CPET alone is unreliable in assessing for ischemia, partly because the positive predictive value is low in children and also because ischemia with AAOCA, when it occurs, is intermittent. In an autopsy series by Basso et al., the authors found that of 6 children who had undergone a maximal CPET in the months prior to SCD, all had normal studies and had been cleared for exercise.[4] In a study by Brothers et al., the authors demonstrated the intermittent nature of ischemia in a patient with undiagnosed interarterial ALCA-R who underwent two CPETs, one of which was positive for ischemia and the other was negative.[8] The CPET may be more useful if coupled with an imaging study, such as a myocardial perfusion scan or stress echocardiogram. As well, an "off protocol" exercise test that mimics the type of exercise in which the patient usually participates may be more likely to reproduce symptomatology and help to demonstrate myocardial ischemia, if present.

Another challenge occurs with determining the ideal management of a patient with AAOCA, especially the asymptomatic patient with ARCA-L. There is significant variability in the treatment and management of patients with AAOCA.[8] Prior to the recent guidelines, patients with both ARCA-L and ALCA-R were either exercise-restricted from competitive sports or were offered surgical repair if the patient wanted to return to athletic participation. However, the recent American Heart Association/American College of Cardiology Scientific Statement[9] distinguishes between the lower risk interarterial ARCA-L and the higher risk interarterial ALCA-R. These guidelines now allow a young patient with ARCA-L who is asymptomatic to be given the option of participating in competitive athletics, after a discussion with the patient and the family about the risks involved with this decision.

In the majority of patients with interarterial, intramural ALCA-R, and symptomatic ARCA-L, the modified unroofing procedure is recommended. While this procedure has a very low mortality rate, generally associated with preoperative ischemia and aborted SCD,[10] there have been several reports of postoperative morbidity.[11,12] In adults, one study found no difference in mortality over a 10-year-period when comparing exercise restriction versus surgery.[13] The long-term morbidity and mortality rate with children and young adults is still unknown. Indeed, to conclude the postoperative risk of SCD is at or below that of the general young athletic population, we would need at minimum 5000 patient years for ALCA-R and 10 000 patient years for ARCA-L. As well, for all these years, patients would need to be between the ages of 12–22 years and all participating in competitive sports.

In summary, most coronary anomalies are benign and do not require surgery or exercise restriction. In the higher risk anomalies, such as interarterial AAOCA, the decision about exercise restriction versus surgical repair is less clear. More data are needed on larger numbers of patients, such as through the multicenter AAOCA registry of the Congenital Heart Surgeons' Society,[14] before we are able to assess treatment efficacy.

CONFLICT OF INTEREST

None.

DISCLOSURES

None.

AUTHOR CONTRIBUTIONS

The author contributed in the concept/design, drafting, critical revision and approval of the article.

REFERENCES

[1] Murphy DA, Roy DL, Sohal M, Chandler BM. Anomalous origin of left main coronary artery from anterior sinus of Valsalva with myocardial infarction. *J Thorac Cardiovasc Surg*. 1978;75:282–285.

[2] Kragel AH, Roberts WC. Anomalous origin of either the right or left main coronary artery from the aorta with subsequent coursing between aorta and pulmonary trunk: analysis of 32 necropsy cases. *Am J Cardiol*. 1988;62:771–777.

[3] Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol*. 1992;20:640–647.

[4] 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;35:1493–1501.

[5] Hill SF, Sheppard MN. A silent cause of sudden cardiac death especially in sport: congenital coronary artery anomalies. *Br J Sports Med*. 2014;48:1151–1156.

[6] Angelini P, Velasco JA, Ott D, Khoshnevis GR. Anomalous coronary artery arising from the opposite sinus: descriptive features and pathophysiologic mechanisms, as documented by intravascular ultrasonography. *J Invasive Cardiol*. 2003;15:507–514.

[7] Taylor AJ, Byers JP, Cheitlin MD, Virmani R. Anomalous right or left coronary artery from the contralateral coronary sinus: high risk abnormalities in the initial coronary artery course and heterogeneous clinical outcomes. *Am Heart J*. 1997;133:428–435.

[8] 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;140:e27–e29.

[9] Van Hare GF, Ackerman MJ, Evangelista JK, et al.; on behalf of the American Heart Association Electrocardiography and Arrhythmias Committee of the Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular and Stroke Nursing, Council on Functional Genomics and Translational Biology, and the American College of Cardiology. Eligibility and disqualification recommendations for competitive athletes with cardiovascular abnormalities: Task Force 4: congenital heart disease: a Scientific Statement from the American Heart Association and American College of Cardiology. *Circulation* 2015;132:e281–e291.

[10] Nguyen AL, Haas F, Evens J, Breur JM. Sudden cardiac death after repair of anomalous origin of left coronary artery from right sinus of Valsalva with an interarterial course: case report and review of the literature. *Neth Heart J*. 2012;20:463–471.

[11] 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;76:589–596.

[12] Wittlieb-Weber CA, Brothers JA. Coronary artery anomalies: current recognition and treatment strategies. Update on recent progress. *Curr Cardiovasc Risk Rep*. 2014;8:395–398.

[13] Krasuski RA, Magyar D, Hart S, et al. Long-term outcome and impact of surgery on adults with coronary arteries originating from the opposite coronary cusp. *Circulation*. 2011;123:154–162.

[14] 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;20:50–58.

How to cite this article: Brothers JA. Introduction to anomalous aortic origin of a coronary artery. *Congenital Heart Disease*. 2017;12:600–602. https://doi.org/10.1111/chd.12497