Received: 27 January 2017 | Revised: 12 May 2017 | Accepted: 4 June 2017

DOI: 10.1111/chd.12511

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Current practices are variable in the evaluation and management of patients with anomalous aortic origin of a coronary artery: Results of a survey

Hitesh Agrawal, MD[1,2] | Carlos M. Mery, MD, MPH[1,3] | Patrick E. Day, BS[4] | S. Kristen Sexson Tejtel, MD, PhD[1,2] | E. Dean McKenzie, MD[1,3,5] | Charles D. Fraser Jr, MD[1,3] | Athar M. Qureshi, MD[1,2] | Silvana Molossi, MD, PhD[1,2]

[1]Coronary Anomalies Program, Texas Children's Hospital, Houston, Texas

[2]The Lillie Frank Abercrombie Section of Cardiology, Baylor College of Medicine, Texas Children's Hospital, Houston, Texas

[3]Division of Congenital Heart Surgery, Michael E. DeBakey Department of Surgery, Texas Children's Hospital, Baylor College of Medicine, Houston, Texas

[4]McGovern Medical School at The University of Texas Health Science Center at Houston, Houston, Texas

[5]Division of Cardiothoracic Surgery, Children's Healthcare of Atlanta, Atlanta, Georgia

Correspondence
Silvana Molossi, Baylor College of Medicine, Texas Children's Hospital, 6621 Fannin Street, WT 19345-C, Houston, TX 77030.
Email: smolossi@bcm.edu

Abstract

Background: Anomalous aortic origin of a coronary artery (AAOCA) is the second leading cause of sudden cardiac death in young athletes in the USA. Long-term outcome data for these patients are lacking to date. There is insufficient knowledge on the best approach to these patients and they are managed in a nonuniform manner.

Methods: An online survey of 15 questions regarding management of AAOCA was sent out to 198 cardiac healthcare providers. The goal was to define gaps in knowledge to justify a dedicated scientific forum for discussion of AAOCA. Descriptive statistics were performed.

Results: A total of 91 providers (46%) completed the survey including pediatric cardiology subspecialists (40%), general pediatric cardiologists (24%), cardiovascular (CV) surgeons (22%), adult cardiologists (10%), nurse practitioners (8%), cardiology fellows (3%) and CV anesthesiologist (1%). Forty-eight percent had been practicing for over 15 years and 28% were in their first 5 years of practice. Fifty-two percent of the providers cared for adults and 93% cared for children/adolescents. Eighty-eight percent were affiliated with an academic institution. All but one provider practiced in the USA, 62% practiced in Texas. Half of participants (50%) were very comfortable managing AAOCA patients and 36% were somewhat comfortable. Providers utilized various imaging tests to confirm the anatomy including computed tomography angiography 88%, cardiac magnetic resonance imaging 70%, cardiac catheterization 60%, echocardiogram 12%, IVUS 2% and myocardial perfusion scan 1%. The majority felt comfortable in counseling the families and felt that depending on the type of lesion these patients should get surgical referral (85%) vs clinical follow up (67%) with exercise restriction (65%).

Conclusion: There is heterogeneity in the way AAOCA patients are currently evaluated and managed. A knowledge gap exists even with participants from academic institutions. Long-term data with a defined approach to management of these patients may help to improve outcomes and prevent unnecessary exercise restriction or surgery.

KEYWORDS

anomalous coronary arteries, congenital heart disease, evaluation and management, survey

1 | BACKGROUND

Anomalous aortic origin of a coronary artery (AAOCA) is a congenital anomaly of the origin or course of a coronary artery that arises from the aorta.[1] The reported prevalence of AAOCA varies depending on diagnostic methods applied: 0.06%–0.9% for anomalous right coronary artery (ARCA), 0.025%–0.15% for anomalous left coronary artery (ALCA) and 0.02%–0.67% for anomalous circumflex coronary artery.[2–4] Importantly, AAOCA especially when the coronary artery originates from the opposite sinus of Valsalva, is the second

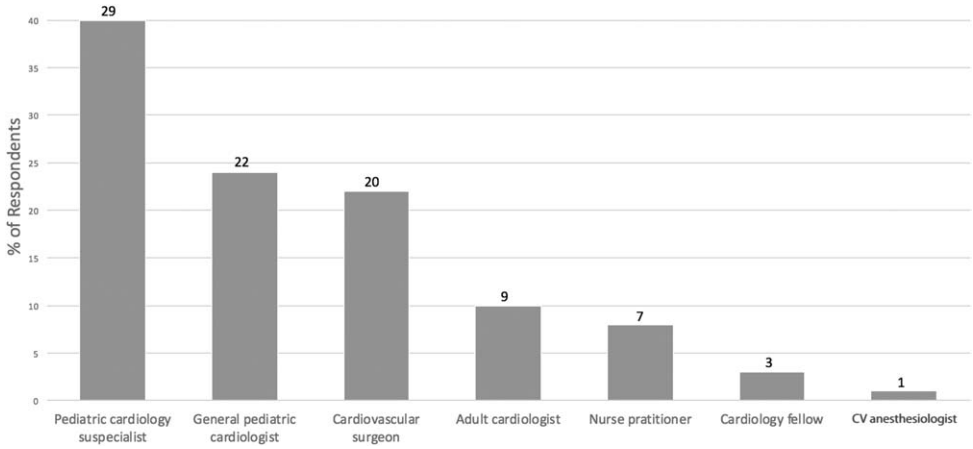

FIGURE 1 Study participants

leading cause of sudden cardiac death (SCD) in young athletes in the USA.[5]

There is lack of clear consensus on the mechanism of ischemia and risk factors leading to SCD in these patients[6] and hence, these patients are evaluated in a non-uniform manner across institutions, even by different providers residing in the same institution.[7] A variety of imaging modalities including echocardiogram, computed tomography angiography (CTA), cardiac magnetic resonance imaging (CMR) and cardiac catheterization have been used to evaluate patients with AAOCA, but a standard imaging protocol is not widely agreed upon.[8,9] Furthermore, even when using the same test such as an echocardiogram, there is poor agreement between institutional reports and imaging core laboratory review for diagnosis of the origin of a coronary artery, intramural and interarterial course.[10]

We believe there is a gap in knowledge, heterogeneous approach, and low comfort level in regards to the evaluation and management of patients with AAOCA among providers. Our goal was to try to define this knowledge gap to potentially justify a dedicated scientific forum for discussion on the best approach to evaluate and manage patients with AAOCA.

2 | METHODS

An online survey of 15 questions regarding evaluation and management of patients with AAOCA was sent out to a total of 198 healthcare providers caring for patients with heart disease at two different times: in June 2013 and in August 2014. The providers were identified based on personal email data base and included professionals and trainees from our own institution as well as others with interest in caring for patients with AAOCA. The survey was modified between the 2013 and 2014 polls: four questions were removed (Q1–4) and four questions were added (Q16–19) on the second survey, but the core eleven questions (Q5–15) were received by all 198 participants (Supporting Information Appendix S1). These new questions related to myocardial bridges and Kawasaki disease. The questions related to myocardial bridges are included in this report while the results relating to Kawasaki disease were not analyzed in this study. Given the anonymity of the responses, respondents may have participated in one or both polls. The questions were structured on a multiple choice format, thus in some questions respondents could select multiple answers that applied (Appendix S1). The survey, sent via email, addressed level of comfort with handling AAOCA patients, surgical intervention, clinical follow up, sports/exercise restrictions, choice of imaging modality and counseling of patients/families. These specific questions were included to capture the tendency of current practices for the diagnosis and treatment of AAOCA. The survey was sent out twice (in both polls) and the providers were instructed to participate only once for each specific poll. Only the questions pertaining to AAOCA were analyzed. Descriptive statistics were performed on the survey results. Frequency counts and percentages were used for categorical variables.

3 | RESULTS

A total of 91 providers (46%) completed the survey between the two polls with respondents including pediatric cardiology subspecialists (40%), general pediatric cardiologists (24%), cardiovascular (CV) surgeons (22%), adult cardiologists (10%), nurse practitioners (8%), cardiology fellows (3%) and CV anesthesiologist (1%) (Figure 1). Respondent providers represented a wide range of clinical experience with 48% having practiced for over 15 years, 24% between 6 and 15 years, and 28% were in their first 5 years of practice (Figure 2). The majority of providers (93%) cared for children and adolescents and 52% cared for adults. Most of the providers (75%) cared for patients with diverse cultural backgrounds and disparate socioeconomic status. Respondents were largely affiliated with academic institutions with 88% having an academic appointment with a medical/university center. All but one provider practiced in the USA; 62% practiced in Texas and out of those, 48% were from Houston.

The survey addressed several questions about the providers' individual practices in regards to imaging and managing AAOCA, as well as their comfort level in managing/counseling the affected families. Half of the participants (50%) were "very comfortable" in assessing and managing AAOCA patients, whereas 46% stated otherwise (36% were "somewhat comfortable" and 10% were "not comfortable") in caring

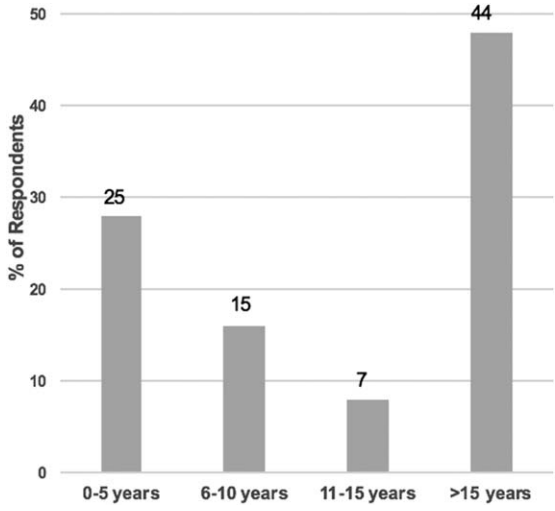

FIGURE 2 Number of years practicing since completion of training

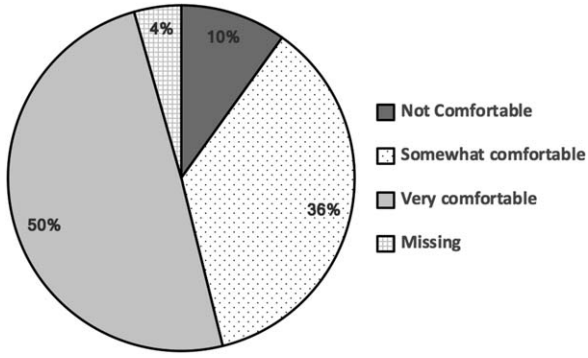

FIGURE 3 Providers' comfort level in assessing and managing patients with anomalous aortic origin of a coronary artery

for these patients (Figure 3). As to the preferred imaging modality to define the coronary anatomy, CTA was most widely used (88%) among the providers, followed by CMR (70%), cardiac catheterization (60%), echocardiogram (12%), intravascular ultrasound imaging (2%) and myocardial perfusion scan (1%) (Figure 4). Approximately two-thirds of respondents (64%) felt comfortable in counseling the families upon confirmation of the diagnosis and 15% reported being uncomfortable in this process. Concerning recommendations for patients with confirmed diagnoses of AAOCA, the respondents felt that, depending on the type of lesion, these patients should get surgical referral (85%) vs clinical follow up (67%) with exercise restriction (65%) (Figures 5 and 6). For those AAOCA patients being followed clinically, about 13% of providers restricted them from intense activity and allowing low to moderate activity, while 3% restricted them from all sports participation (Figure 6).

In regards to the management of patients with myocardial bridges, 47% of respondents had diagnosed <10 such cases in their career and 31% had never diagnosed a patient with a myocardial bridge. A majority of respondents (66%) were not totally comfortable in evaluating and managing patients with myocardial bridges.

4 | DISCUSSION

Our results indicate a heterogeneity of approaches in the evaluation and management of patients with AAOCA despite most participants belonging to academic institutions. In addition, there is an added layer of heterogeneity in the diagnoses bucket of AAOCA, including whether the anomalous coronary is left or right and arising from the opposite sinus, definition of the anomalous course (ie, intramural, interarterial) or the presence of myocardial bridge. Due to inherent limitations of a survey, these details were not available for analyses. Corroborating previous study by Brothers et al.,[7] this heterogeneity indicates a lack of consensus on all stages of the care in patients with AAOCA and the need for defined standard guidelines in both diagnosis and treatment. Until formal guidelines are available and standard of care is established in this population, there is a need to continually educate providers with emerging data. In our study, even more pronounced were the findings regarding the evaluation and management of patients with myocardial bridges, a lesser common diagnosis in children.

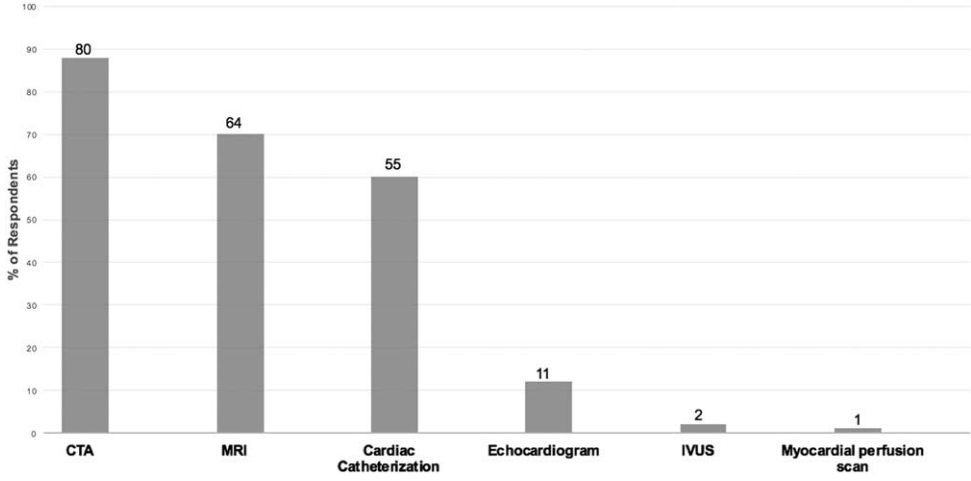

FIGURE 4 Modes of imaging utilized to confirm the anomalous coronary artery anatomy

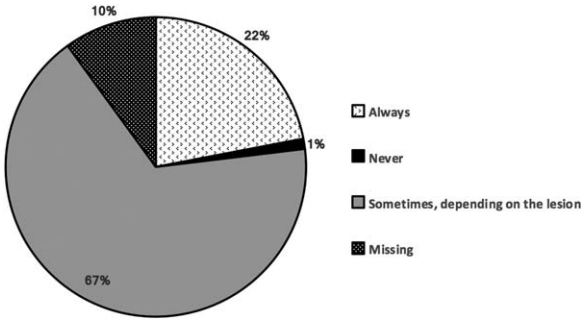

FIGURE 5 Frequency of clinical follow-up only recommended by providers

We have demonstrated in this survey that there is great variability in the selection of preferred tests, both invasive and non-invasive, for confirmation of diagnosis and anatomic details of the anomalous coronary artery. If such practice were to continue, many patients would undergo a multitude of unnecessary tests potentially leading to overuse of resources. Given that a significant proportion of providers considered themselves "not comfortable/somewhat comfortable" in caring for patients with AAOCA (46%) and myocardial bridges (66%), there is clearly a need for data-driven guidance in the evaluation and management of these patients.

The long-term natural and unnatural history of patients with AAOCA remains to be determined.[11–13] AAOCA is a rare condition, necessitating multicenter collaboration for investigators to unravel the natural treated and untreated history of this condition. Early efforts included the establishment of the Congenital Heart Surgeons' Society AAOCA Registry in 2009.[14] This registry currently consists of retrospective and prospective data from 39 North American member institutions. Although this is a step in the right direction, patients continue to be managed in a heterogeneous manner across institutions. This is a major hurdle/confounder to making reliable conclusions from these data, although certainly contributing to our understanding of AAOCA.[1,8,15]

This heterogeneity in evaluating and managing patients with AAOCA and the uncertainty of outcomes led to the development of a dedicated Coronary Anomalies Program (CAP) at our institution. In December 2012, Texas Children's Hospital created a multidisciplinary CAP which includes cardiologists, congenital heart surgeons, cardiovascular radiologists, outcomes and research staff, with the development of a clinical algorithm based on available evidence.[1] Since its inception, all patients with AAOCA and myocardial bridges at our institution have been evaluated and managed in a uniform manner. Adoption of a standardized approach in individual institutions will foster the acquisition of data, a paramount step to making reliable conclusions and to determine the best strategy to improve outcomes for this population of patients, likely contributing to the development of national guidelines.

The recent AHA/ACC scientific statement for sports participation in children and young adults with AAOCA reflect some advancements in this field.[16] It differentiates between higher risk interarterial anomalous left coronary artery (ALCA) and lower risk interarterial anomalous right coronary artery (ARCA). For asymptomatic patients with ARCA and a negative exercise stress test, the recommendation is to allow full activity without any intervention,[16] although with limited data this approach still remains controversial as to its long-term effect, especially in patients with ostial abnormalities and a long intramural course.

The evidence is continually growing in this field and there is a need to regularly update providers with the knowledge being accrued. Following the inception of our CAP, a multidisciplinary conference on coronary artery anomalies was undertaken in 2013, 2014 and 2016, where speakers from several leading institutions provided a dedicated forum to discuss the diagnosis and management of such patients. It continues to be an ever expanding forum for providers caring for such patients and aiming to keep them abreast of the developments in this area. A panel discussion with families affected by AAOCA during these meetings brought special attention to the psychosocial needs of patients as well as their parents and siblings. As in any other field of medicine, quality of life measurement should be standard of care for these patients and families.[17] Improved counseling and measurement of exercise performance over time will help to improve the care and well-being of these patients.

While we believe this study has contributed to our understanding of how providers evaluate and manage this patient population, this survey has several limitations. Firstly, it was sent to almost exclusively US providers (99%) and most of those providers were from Texas, so our results are representative of this specific geographic sample, in addition to potentially having had the same providers answering to both polls. Secondly, most providers (88%) belonged to academic institutions, and likely to a large part our own, which may also bias the results. The anonymity of responders precludes identification. Finally, a variety of providers participated in the study including pediatric cardiology subspecialists, general pediatric cardiologists, CV surgeons, adult cardiologists (10%), nurse

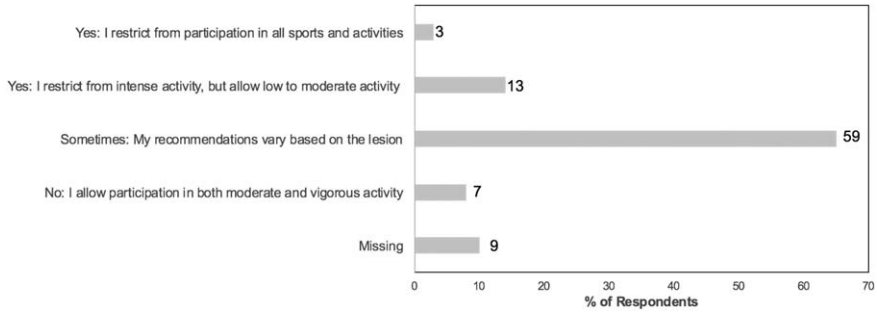

FIGURE 6 Restriction on sports participation when clinical follow up is recommended

practitioners and cardiology fellows, which may have skewed the data accordingly to their expertise and clinical area.

5 | CONCLUSIONS

Our study demonstrates, in a limited sample of healthcare providers, heterogeneity in the way AAOCA patients are currently evaluated and managed by CV specialists. In addition, this study highlights the low comfort level many providers experience in the evaluation and management of patients with AAOCA, a leading cause of SCD in young athletes in the USA. A knowledge gap exists even in providers belonging to academic institutions. Long-term data with a defined approach to evaluation and management of these patients will help to improve outcomes and hopefully prevent unnecessary exercise restriction or surgery. We are currently working on creating a database which will house detailed clinical information on patients diagnosed with anomalous aortic origin of a coronary artery and/or myocardial bridge at Texas Children's Hospital. Once this is complete, the intent is to propose to other institutions to contribute with, and share, prospective clinical, imaging, and outcomes data.

ACKNOWLEDGMENT

There is no funding information to disclose.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

All the listed authors have made significant contribution in the preparation and review of the manuscript.

REFERENCES

[1] Mery CM, Lawrence SM, Krishnamurthy R, et al. Anomalous aortic origin of a coronary artery: toward a standardized approach. *Semin Thorac Cardiovasc Surg*. 2014;26:110–122.

[2] Prakken NH, Cramer MJ, Olimulder MA, Agostoni P, Mali WP, Velthuis BK. Screening for proximal coronary artery anomalies with 3-dimensional MR coronary angiography. *Int J Cardiovasc Imaging*. 2010;26:701–710.

[3] Pelliccia A, Spataro A, Maron BJ. Prospective echocardiographic screening for coronary artery anomalies in 1,360 elite competitive athletes. *J Am Coll Cardiol*. 1993;72:978–979.

[4] Angelini P, Shah NR, Uribe CE, et al. Novel MRI based screening protocol to identify adolescents at high risk of sudden cardiac death. *J Am Coll Cardiol*. 2013;61:E1621.

[5] 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;119:1085–1092.

[6] Kaushal S, Backer CL, Popescu AR, et al. Intramural coronary length correlates with symptoms in patients with anomalous aortic origin of the coronary artery. *Ann Thorac Surg*. 2011;92:986–991.

[7] Brothers J, Gaynor JW, Paridon S, Lorber R, Jacobs M. Anomalous aortic origin of a coronary artery with an interarterial course: understanding current management strategies in children and young adults. *Pediatr Cardiol*. 2009;30:911–921.

[8] 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.

[9] Angelini P, Flamm SD. Newer concepts for imaging anomalous aortic origin of the coronary arteries in adults. *Cathet Cardiovasc Intervent*. 2007;69:942–954.

[10] Lorber R, Srivastava S, Wilder TJ, et al. Anomalous aortic origin of coronary arteries in the young: echocardiographic evaluation with surgical correlation. *JACC Cardiovasc Imaging*. 2015;8:1239–1249.

[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;76:589–595.

[12] Sharma V, Burkhart HM, Dearani JA, et al. Surgical unroofing of anomalous aortic origin of a coronary artery: a single-center experience. *Ann Thorac Surg*. 2014;98:941–945.

[13] Davies JE, Burkhart HM, Dearani JA, et al. Surgical management of anomalous aortic origin of a coronary artery. *Ann Thorac Surg*. 2009;88:844–848.

[14] Brothers JA, Gaynor JW, Jacobs JP, Caldarone C, Jegatheeswaran A, Jacobs ML, et al. The registry of anomalous aortic origin of the coronary artery of The Congenital Heart Surgeons' Society. *Cardiol Young*. 2010;20:50–58.

[15] Angelini P. Coronary artery anomalies – current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. *Tex Heart Inst J*. 2002;29:271–278.

[16] Van Hare GF, Ackerman MJ, Evangelista J-AK, et al. 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.

[17] Sing AC, Tsaur S, Paridon SM, Brothers JA. Quality of life and exercise performance in unoperated children with anomalous aortic origin of a coronary artery from the opposite sinus of valsalva. *Cardiol Young*. 2016;26:1–10.

SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

APPENDIX S1. Questionnaire: AAOCA needs assessment of providers

How to cite this article: Agrawal H, Mery CM, Day PE, et al. Current practices are variable in the evaluation and management of patients with anomalous aortic origin of a coronary artery: Results of a survey. *Congenital Heart Disease*. 2017;12:610–614. https://doi.org/10.1111/chd.12511