Received: 11 May 2017 | Accepted: 20 May 2017

DOI: 10.1111/chd.12493

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Decision making in anomalous aortic origin of a coronary artery

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Abstract

Anomalous aortic origin of a coronary artery (AAOCA) is the second most common cause of sudden cardiac death (SCD) in young athletes in United States. The exact pathophysiological mechanisms of SCD are unknown. There is lack of long-term outcome data on repaired and unrepaired AAOCA and our current risk stratification scheme for these patients is suboptimal. These patients are evaluated in a nonuniform manner across institutions in United States, and even by different providers residing in the same institution. The main objective of this article is to use what is known and unknown about this disease and to provide a possible framework that can help workup and manage patients with AAOCA in a more consistent fashion.

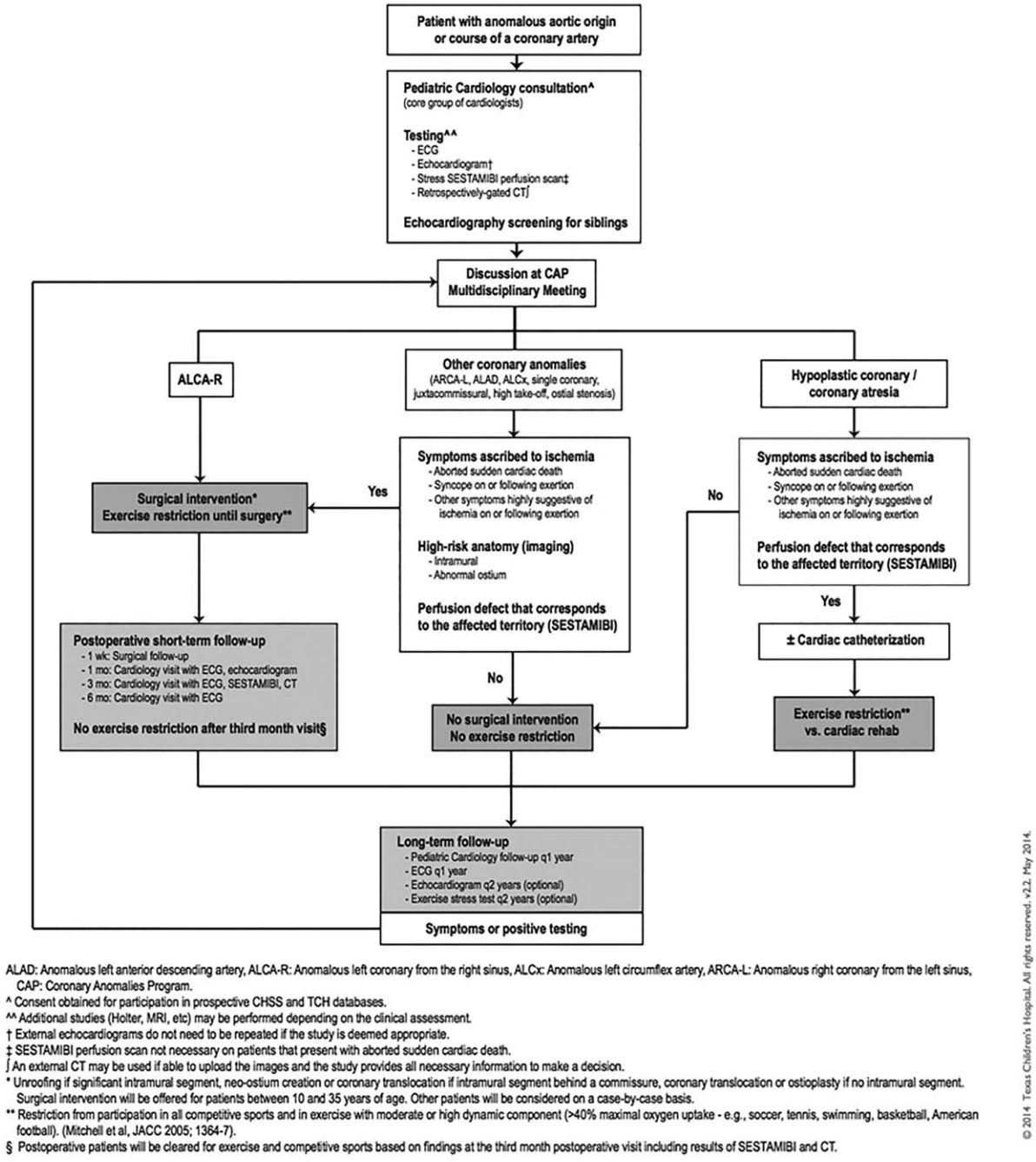
KEYWORDS

anomalous aortic origin of a coronary artery, coronary anomalies, sudden cardiac death

Anomalous aortic origin of a coronary artery (AAOCA) originating from the opposite sinus of Valsalva represents the second most common cause of sudden cardiac death (SCD) in young athletes in the United States.[1] The best management strategy for patients with AAOCA is controversial mainly due to uncertainties surrounding the exact prevalence of the disease, the actual risk of SCD, the risk profile for the different types of AAOCA, and the effect of the available management strategies on decreasing that risk.

Data on AAOCA is scant. However, based on several reports of the literature, some of the following conclusions can be made:

- The exact prevalence of AAOCA is largely unknown. Studies of different populations (including echocardiography,[2] cross-sectional imaging,[3,4] cardiac catheterization,[5,6] and autopsy[7]) have calculated the prevalence to be between 0.1% and 1%. Preliminary results of a recent screening study using MRI on 1836 middle school students show a prevalence of 0.7%.[8] The same studies suggest that an anomalous right coronary artery (ARCA) from the left sinus of Valsalva is approximately 6 times more frequent than an anomalous left coronary artery (ALCA) from the right sinus of Valsalva.
- Despite the higher prevalence of ARCA than ALCA, ALCA patients are more prevalent in series of SCD (4:1 to 5:1),[9,10] leading to the conclusion that ALCA is associated with a higher risk of SCD than ARCA.
- Extrapolating the results of prevalence studies and epidemiological series on SCD in high school and college athletes, one can calculate that the annual risk of SCD in young athletes with AAOCA is probably between 0.08% and 0.26% for ALCA and 0.003% and 0.01% for ARCA if the prevalence is 0.7% and between 0.58% and 1.79% for ALCA and 0.02% and 0.08% for ARCA if the prevalence is 0.1%.
- Most events of SCD occur during or immediately after vigorous physical activity with a high dynamic component.[9] Patients with SCD may have exercised before multiple times at higher levels of exertion without previous problems.
- On retrospect, approximately 50% of patients that suffer SCD had complained of symptoms before.[9,11] Several patients have had a negative exercise stress test several months prior to the SCD event.[9]
- SCD is rare in patients <10 or >30 years of age.[10]
- The exact pathophysiological mechanism for SCD in AAOCA is unknown. Theories include intermittent compression of an intramural segment of the anomalous coronary as it travels within the wall of the aorta (a large proportion of these patients have an intramural segment), compression of the coronary as it travels between the aorta and the pulmonary artery, and morphological alterations of the ostium of the anomalous vessel (eg, a slit-like ostium or ostial stenosis).
- Intravascular ultrasound studies have shown that the intramural segment of anomalous coronaries is 50%–70% smaller in circumference than the distal segment of the same coronary artery due to lateral compression and that the compression worsens with fluids and dobutamine to simulate exercise conditions.[12]



ALAD: Anomalous left anterior descending artery, 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.
^ Consent obtained for participation in prospective CHSS and TCH databases.
^^ Additional studies (Holter, MRI, etc) may be performed depending on the clinical assessment.
† External echocardiograms do not need to be repeated if the study is deemed appropriate.
‡ SESTAMIBI perfusion scan not necessary on patients that present with aborted sudden cardiac death.
∫ An external CT may be used if able to upload the images and the study provides all necessary information to make a decision.
* Unroofing if significant intramural segment, neo-ostium creation or coronary translocation if intramural segment behind a commissure, coronary translocation or ostioplasty if no intramural segment. Surgical intervention will be offered for patients between 10 and 35 years of age. Other patients will be considered on a case-by-case basis.
** Restriction from participation in all competitive sports and in exercise with moderate or high dynamic component (>40% maximal oxygen uptake - e.g., soccer, tennis, swimming, basketball, American football). (Mitchell et al, JACC 2005; 1364-7).
§ Postoperative patients will be cleared for exercise and competitive sports based on findings at the third month postoperative visit including results of SESTAMIBI and CT.

FIGURE 1 Algorithm used by the Texas Children's Hospital Coronary Anomalies Program for workup and management of patients with an abnormal origin or course of a coronary artery. © 2014 Texas Children's Hospital (reprinted with permission)

Given all these uncertainties and in an effort to provide a more consistent way to manage patients with AAOCA, the Coronary Anomalies Program (CAP) at Texas Children's Hospital was created in December 2012 by a multidisciplinary team of cardiologists, radiologists, congenital heart surgeons, and researchers. As part of the CAP, a workup and management algorithm was developed based on the limited data available and consensus discussions. As experience has accrued over the past few years, the algorithm has been revised appropriately. Figure 1 shows the most currently used algorithm for workup and management of patients with AAOCA.

All patients with coronary anomalies are seen by one of two cardiologists that are part of the CAP. Patients undergo a series of standardized tests including retrospective electrographically gated computerized tomography to define the location and morphology of the ostium and the presence and length of intramurality (based on the morphology of the proximal coronary and the presence or absence of pericoronary fat), and treadmill exercise stress test with nuclear perfusion imaging (and more recently stress magnetic resonance imaging) to assess myocardial perfusion at rest and upon stress. All patients are then presented in a dedicated multidisciplinary monthly meeting to analyze their data.

Patients that are believed to have a higher risk lesion amenable to surgical intervention are offered surgery with exercise restriction until after 3 months postoperatively (provided all testing is normal at the time). Patients that are believed to have a lower risk lesion are cleared for exercise. All patients are entered in a longitudinal database and followed closely. Since the inception of the program, two thirds of the patients seen were diagnosed with ARCA and one fourth of all patients underwent surgical intervention, with unroofing being the most common procedure performed. All patients that have undergone surgical intervention have been cleared for exercise 3 months postoperatively.

Even though the best workup and management strategy for patients with AAOCA is still unclear, the creation of a dedicated multidisciplinary program has allowed the use of a more consistent approach for these patients. Careful longitudinal data collection and analysis and participation in multi-institutional registries such as the one created by the Congenital Heart Surgeons Society[13] will hopefully provide more insight in the future and allow us to have a more informed and data-driven management strategy for these patients.

CONFLICT OF INTEREST

None.

DISCLOSURES

None.

AUTHOR CONTRIBUTIONS

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

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How to cite this article: Mery CM. Decision making in anomalous aortic origin of a coronary artery. *Congenital Heart Disease.* 2017;12:630–632. https://doi.org/10.1111/chd.12493