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Clinical evaluation of anomalous aortic origin of a coronary artery (AAOCA)

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Abstract

The clinical evaluation of patients with an anomalous aortic origin of a coronary artery (AAOCA), a congenital abnormality of the origin or course of a coronary artery that arises from the aorta, is challenging given its first presentation being sudden cardiac arrest in about half of the patients. Symptoms of chest pain, shortness of breath and syncope during exertion should be of concern in evaluating young athletes and nonathletes. The lack of abnormal signs on the physical exam and electrocardiogram further adds to the difficulty in establishing the diagnosis. Additional imaging with echocardiography, computed tomography angiography and/or cardiac magnetic resonance imaging (MRI) is often needed and establishes the diagnosis. High-risk lesions include origin of the coronary artery from the opposite sinus of Valsalva, intramural course and ostial abnormalities (stenosis, hypoplasia). Functional studies should be performed to assess myocardial perfusion at rest and during stress, such as nuclear imaging, stress echocardiography and stress cardiac MRI.

KEYWORDS

anomalous aortic origin of a coronary artery, congenital heart disease, coronary anomalies, myocardial perfusion, sudden cardiac death

1 | INTRODUCTION

The clinical evaluation of patients with an anomalous aortic origin of a coronary artery (AAOCA), a congenital abnormality of the origin or course of a coronary artery that arises from the aorta, is not infrequently a difficult task. This condition, especially when the anomalous vessel originates from the opposite sinus of Valsalva, is associated with sudden cardiac death (SCD) accounting for 14%–17% of all cardiac-related deaths in young athletes.[1–3] An increasing number of children and adolescents are being diagnosed with AAOCA following routine pre-participation screening, presence of a murmur or an abnormal ECG. Further evaluation, without being driven by concerns with occurrence of symptoms, then leads to additional imaging and the diagnosis of AAOCA.

It is known that about 75% of SCD is related to a cardiovascular etiology, usually during or following exercise activity and most commonly associated with dynamic exercise.[1,4,5] Several pathophysiologic mechanisms have been postulated for the occurrence of sudden cardiac arrest (SCA) in patients with AAOCA. These include occlusion and/or compression of the anomalous coronary artery (intramural segment, interarterial course) and ostial abnormalities (slit-like and stenotic ostium), particularly during exercise, leading to myocardial ischemia and development of ventricular arrhythmia.[6–8] However, it is not understood why these patients often performed the same level and intensity of exercise for many years without symptoms prior to an SCA/SCD event and what leads to the occurrence of SCA/SCD at rest in both athletes and nonathletes.

2 | CLINICAL PRESENTATION

The clinical presentation of AAOCA is variable, symptoms may not be present in a large number of patients, and an episode of SCA or SCD might be the initial event in a few. About half of the patients with AAOCA are asymptomatic and the other half may present with chest pain (on exertion or at rest), palpitations, shortness of breath, dizziness or syncope on exertion.[2–4,9–11] These symptoms are quite common in the outpatient pediatric cardiology practice and this young population is the one at risk for sudden events, making the validation of symptoms as it relates to the diagnosis even more complex.

Basso et al.[9] reported only 10 (36%) of 27 cases presenting with SCD (23 anomalous left and 4 anomalous right coronary artery from the opposite sinus of Valsalva) had symptoms prior to the event

including syncope, chest pain and palpitations. All cases had an acute angle take off and a slit-like ostium. Eckart et al.[11] reported 11 (52%) of 21 military recruits with AAOCA suffering SCD had previous symptoms of syncope, chest pain and dyspnea.

Chest pain, however, is a common complaint accounting for up to 5% of new consultations in the pediatric cardiology clinic, up to 13% of EC visits as a whole and up to 19% of new EC visits.[12] Saleeb et al.[13] published 3700 cases of chest pain seen in the pediatric cardiology clinic over 10 years period with causes attributed to idiopathic in 52%, musculoskeletal in 36%, respiratory in 7%, gastrointestinal in 3% and only 1% of cardiac etiology (37 cases: 16 SVT, 10 pericarditis). A specific cardiac diagnosis was identified in 9 cases (0.24%), 4 with myocarditis, 3 with AAOCA, 2 with hypertrophic cardiomyopathy and 1 with dilated cardiomyopathy. Over 18 000 patients years of follow up in this cohort there was no missed diagnosis leading to SCD. Syncope is also a common complaint between 15 and 19 years of age, especially females and the etiology is neurocardiogenic/neurologic in ~95% of patients.[14,15] It is estimated that 3%-5% of patients will have a significant cardiac diagnosis, and mostly will be related to channelopathies.[15]

In 90 patients (mean age 11.8 ± 4.2, range of 0.08–19 years) evaluated at the Coronary Anomalies Program at Texas Children's Hospital created in December 2012, to December 2014, 51% were asymptomatic, 29% had chest pain (54% on exertion, 23% on both exertion and at rest and 23% only at rest), 15% had syncope (57% on exertion), and 3.3% presented with SCA.[16] Brothers and Harris[17] reported limited episode of exercise-induced ventricular tachycardia in a patient with ARCA and this also observed in our series[16] interestingly with no report of palpitations.

3 | FUNCTIONAL ASSESSMENT OF MYOCARDIAL ISCHEMIA

Functional assessment of myocardial ischemia is routinely performed to evaluate AAOCA patients with and without symptoms, though the validity is yet to be clearly defined given reports of both false-positive and false-negative results. Exercise stress test is universally used in the evaluation of these patients. In the study, analyzing 27 cases of SCD in athletes with AAOCA by Basso et al.,[9] a maximal exercise stress test (EST) had been performed and deemed normal in six patients 6–18 months prior to their death. In their review of the literature, 18 patients with AAOCA that had EST findings reported were found, with positive findings in only 4 (22%) patients, including 2 patients that were symptomatic. All five patients that died from AAOCA had a negative EST within 6 months prior to death.[9] In the study by Brothers et al.,[18] only 1 (6%) of 16 patients that underwent surgical intervention for AAOCA had a positive EST prior to surgery. In the 90 patients evaluated in our program thus far, we have not observed a single EST that was definitive for myocardial ischemia.

Nuclear stress perfusion (NSP) imaging is also often utilized in these patients to evaluate myocardial perfusion and both false-positive and false-negative results question its reliability to identify true myocardial ischemia. Brothers et al.[18] reported 10 patients undergoing NSP imaging following surgical repair for AAOCA. Of these, three had perfusion defects identified, though no corresponding preoperative evaluation was performed for comparison.

Pharmacologic stress testing by cardiac magnetic resonance imaging (MRI) has been shown to be superior to nuclear techniques in the adult population in assessing myocardial perfusion,[19] though its use in the pediatric population and especially in AAOCA patients is scarce at this time. This seems to be a promising technique that may help in the future to identify myocardial perfusion defects in select patients and contribute to risk stratification.

In our Coronary Anomalies Program at Texas Children's Hospital, we have adopted NSP imaging following treadmill exercise test with myocardial oxygen consumption measurement to assess myocardial perfusion following detailed anatomy by chest tomography angiogram. We have started to develop a specific protocol to assess myocardial perfusion with stress cardiac MRI. We will compare with NSP and hope to determine sensitivity and specificity for both modalities. Cardiac catheterization has been performed in a limited number of cases and we believe this can be useful, specially with determination of coronary fractional flow reserve and also utilizing intravascular ultrasound.

4 | CONCLUSIONS

The optimal strategy to evaluate patients with AAOCA is yet to be defined. Several factors, including the commonality of ominous symptoms in the pediatric population and the infrequent signs of myocardial ischemia prior to a sudden event, continue to challenge providers and care takers to identify those patients who are at risk of SCA/SCD in the general population. The future looks promising as growing efforts are occurring at the national level to obtain more objective data, particularly related to defining the anatomic types that confer high risk[20,21] and the creation of registries with long-term longitudinal follow-up.[22]

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CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

Concept/design: Molossi, Agrawal
Drafting: Molossi, Agrawal
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