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INTERMEDIATE

CASE REPORT: CLINICAL CASE SERIES

Case Series of Coronary Artery Anomalies in Athletes

Challenges in Clinical Management and Sports Eligibility

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ABSTRACT

Coronary artery anomalies include a spectrum of pathologic changes associated with sudden cardiac death in athletes. We highlight the inherent challenges in risk stratification and management of athletes with coronary artery anomalies by presenting 3 cases, each with distinct pathologic coronary anatomy and clinical management decisions. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:1074–1079) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Coronary artery anomalies (CAAs) include numerous variants, most of which are benign and incidental.[1] However, rare malignant CAAs, such as an anomalous interarterial left coronary artery, are associated with sudden cardiac death (SCD) in young athletes.[2] The symptoms may be variable, and cardiac screening techniques lack sensitivity and specificity for the detection of pathologic CAAs. Evidence-based practices for the management of most pathologic CAAs, including sports eligibility, are lacking.[3,4] We present 3 cases of young athletes with CAAs, highlighting the heterogeneity in anatomical variation and challenges inherent within risk stratification and clinical management strategies.

LEARNING OBJECTIVES

- To review the clinical evaluation of pathologic coronary artery anomalies in athletes.
- To highlight shared decision making when counseling athletes with coronary artery anomalies.

CASE 1

A 19-year-old female collegiate basketball player with no prior cardiac symptoms was referred to sports cardiology after a syncopal episode at practice. She had been followed by cardiology for the previous 3 years because of an anomalous interarterial right coronary artery (ARCA) that had been identified incidentally by a screening echocardiogram before her participation in sports (Figure 1). She had previously been risk stratified with annual exercise testing. With this exertional syncopal event, she had been running suicide intervals and remembered a brief period of lightheadedness before loss of consciousness. She regained consciousness in several seconds.

A coronary computed tomography angiogram (CCTA) demonstrated an interarterial ARCA with malignant anatomical features, including a slitlike orifice and severely narrowed proximal intramural segment (Figure 2). Dobutamine stress cardiac magnetic resonance (CMR) did not demonstrate evidence of myocardial fibrosis or ischemia, although the

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maximum heart rate achieved was only 170 beats/min.

Although it was uncertain whether her syncopal episode was attributed to the ARCA, imaging suggested malignant CAA anatomy.[1] Coupled with exertional syncope, surgical unroofing of the ARCA was recommended.[4] The patient has since undergone uncomplicated surgery and awaits her return to training after 3 months of abstinence from sports.[3]

CASE 2

A 16-year-old male athlete playing American-style football was referred to sports cardiology after discovery of an ARCA and left anterior descending artery (LAD) myocardial bridge. These CAAs were simultaneously discovered by CCTA after 3 months of exertional chest tightness.

Exercise testing revealed ST depressions at peak exertion associated with chest pain (**Figure 3**). Subsequent dobutamine stress CMR demonstrated perfusion defects in the anteroseptal, apical lateral, and inferolateral walls at a maximum heart rate of 134 beats/min (**Figure 4**), without evidence of myocardial scar. Coronary angiography demonstrated a wrap-around LAD and long mid-LAD myocardial bridge with an instantaneous wave-free ratio of 0.86 that normalized to 1.0 on proximal pullback. Instantaneous wave-free ratio of the ARCA was 1.0 (**Figure 5, Video 1**).

In this case, high-risk proximal anatomical ARCA features[3,4] were absent. Although myocardial bridges are commonly clinically inconsequential,[5] there was objective evidence of ischemia in the LAD territory, implicating this CAA as the underlying cause of symptoms. After extensive discussions, the athlete and his parents preferred a conservative approach and avoidance of surgery. A strategy of pre-exercise short-acting β-blocker agent was trialed, titrated, and well tolerated.[6] Repeated exercise testing and dobutamine stress CMR while the patient was using the β-blocker agent yielded normal results and did not induce symptoms. By use of a shared decision making (SDM) approach, the athlete returned to sport with this medical strategy in place. Importantly, his school and athletic trainers were engaged with SDM, supporting his participation and affirming that appropriate emergency action plans were in place. He continued with annual exercise and functional testing throughout the remainder of his high school athletic career.

CASE 3

A 17-year-old male hockey athlete, previously evaluated by cardiology, was referred to sports cardiology after 2 months of recently resolved chest pain. The chest pain was a tight substernal sensation occasionally occurring at peak exertion but also several times per week at rest. The coronary ostia could not be identified by echocardiography; thus, CCTA was obtained and demonstrated a hypoplastic LCA (**Figure 6**).

ABBREVIATIONS AND ACRONYMS

ARCA = anomalous right coronary artery from the left sinus of Valsalva

CAA = coronary artery anomaly

CCTA = coronary computed tomography angiography

CMR = cardiac magnetic resonance

LAD = left anterior descending coronary artery

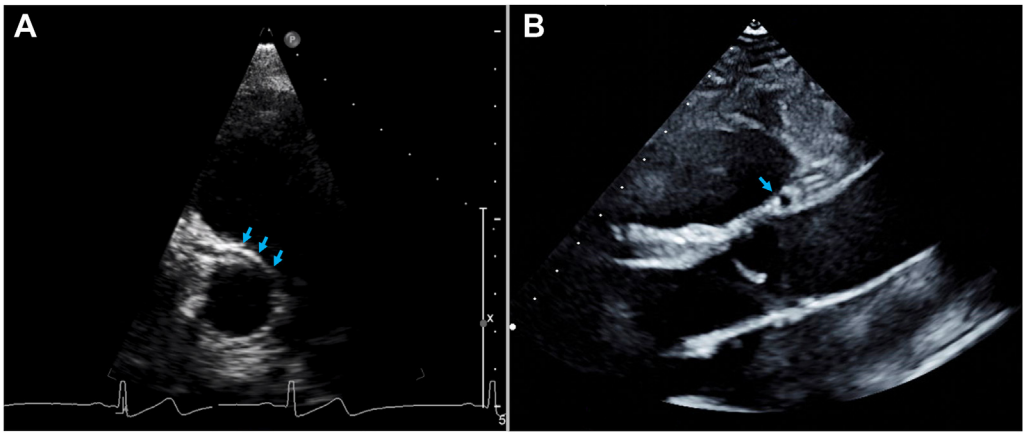
LCA = left coronary artery

RCA = right coronary artery

SCD = sudden cardiac death

SDM = shared decision making

FIGURE 1 Transthoracic Echocardiography of an Anomalous Coronary Origin



(A) Parasternal short-axis view at the aortic root suggesting an anomalous right coronary artery from the left sinus of Valsalva. **(B)** Parasternal long-axis view with "ring sign" at the level of the sinotubular junction suggestive of an anomalous right coronary artery.

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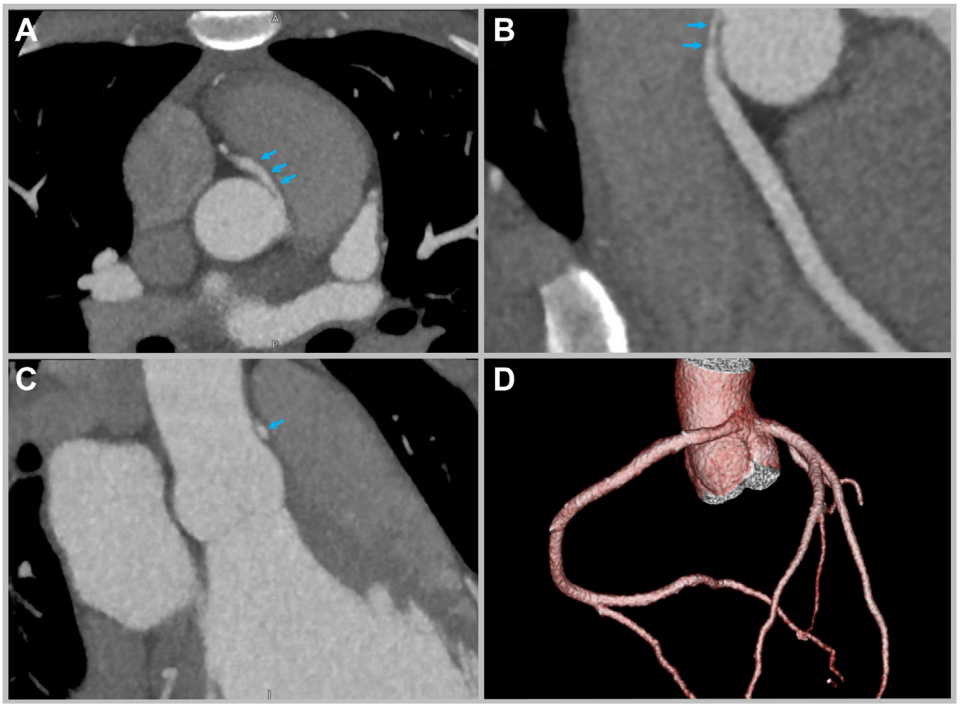
FIGURE 2 Computed Tomography Angiography



(A) Axial view of the anomalous right coronary artery originating from the left coronary cusp with an acute angle takeoff. **(B)** Intramural and extremely narrow ostium and proximal portion of the anomalous right coronary artery. **(C)** Cross-sectional view of the proximal anomalous right coronary artery demonstrating slitlike anatomy. **(D)** 3-Dimensional reconstruction demonstrating anomalous right coronary artery from the left coronary cusp.

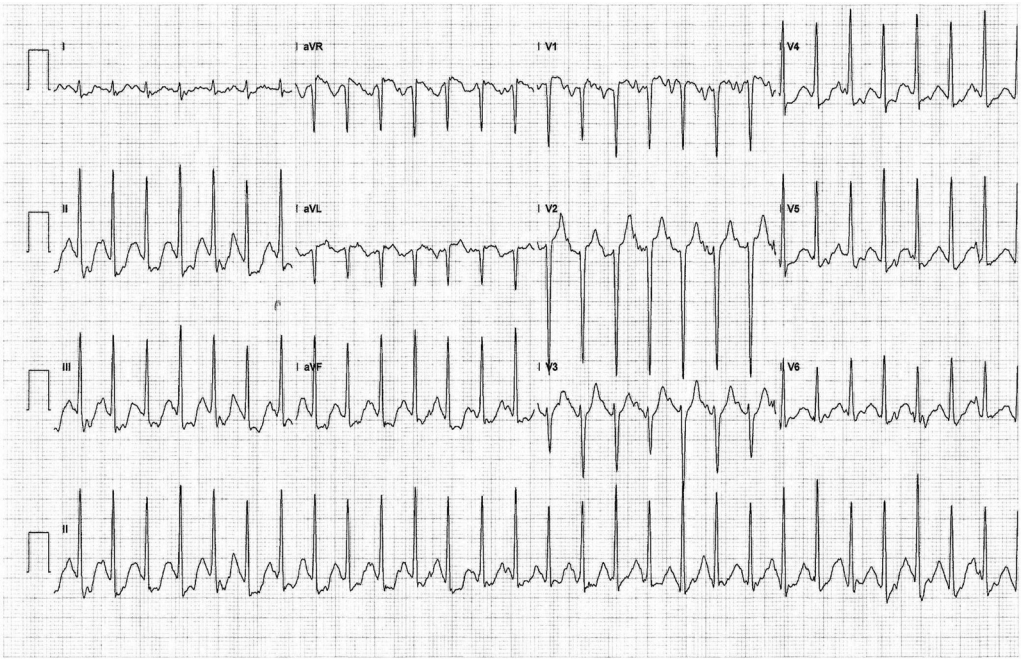
Exercise testing did not elicit chest pain or ECG abnormalities, and dobutamine stress CMR did not demonstrate ischemia. Coronary angiography confirmed a diminutive LCA but also a large, dominant RCA with extensive septal collateralization of the anterior myocardial wall (Figure 7).

Hypoplastic LCA is a rare CAA that can be associated with ischemia and SCD.[4] In this case, the chest pain was described with noncardiac features and recent temporal cessation. Coupled with the extensive collateral coronary circulation and lack of high-risk stress testing features, it was believed that there was room for a nuanced SDM approach for sports eligibility. After extensive discussions detailing his uncertain risk of a cardiac event with intense activity, he and his family still desired a return to hockey. With an emergency action plan in place in coordination with his school and trainers, he has returned to sport without recurrence of symptoms and is followed annually with surveillance exercise testing.

DISCUSSION

These 3 cases highlight numerous clinical challenges associated with CAAs in athletes. First, malignant CAAs are rare variants associated with an increased risk of SCD during exercise. They do not reliably present with exertional syncope or chest pain, nor are they detectable by standard preparticipation screening. Second, management is complicated because of the wide range of anatomical variants and the uncertain risks associated with exercise training for many pathologic CAA phenotypes. Finally, CAAs diagnosed in athletes with exertional symptoms and malignant CAA phenotypes discovered in asymptomatic athletes require intensive risk stratification with imaging, functional testing, and consideration of referral to a dedicated coronary anomalies program. With the lack of robust outcomes data, however, clinical management options and sports eligibility often require an individualized SDM approach.

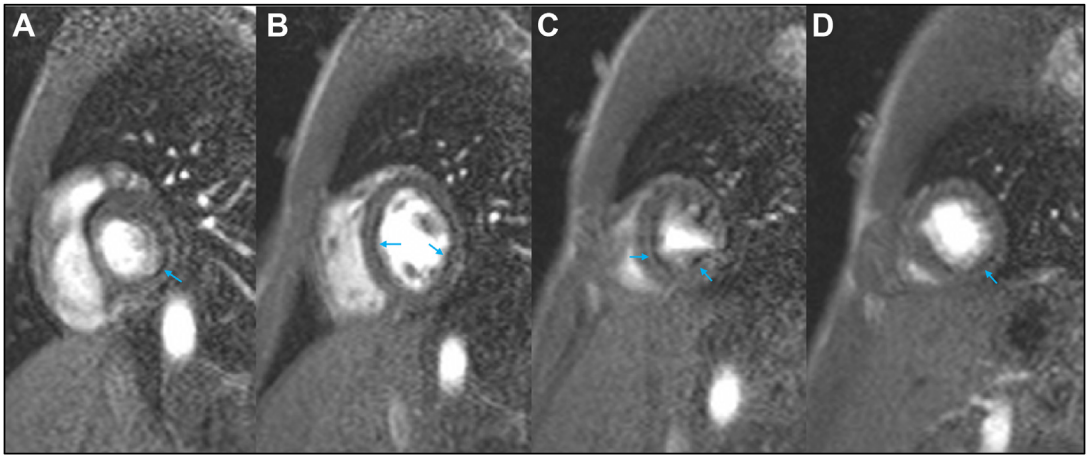
FIGURE 3 Stress Electrocardiogram



Stress electrocardiogram demonstrating inferior ST-segment depressions at a heart rate of 175 beats/min.

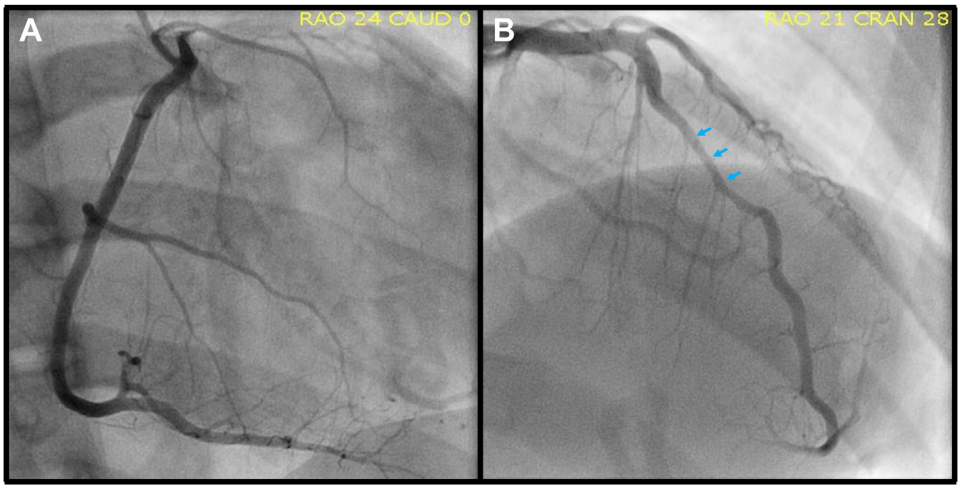
In the cases presented here, risk stratification with imaging and functional ischemic assessments were key in estimating qualitative risk. CCTA is the gold standard imaging modality for anatomical CAA evaluation because of its ability to characterize malignant features, including a slit-like orifice, intramural course, or long stenotic area.[1] CMR also carries a Class I indication for anatomical evaluation of CAAs

FIGURE 4 Stress Magnetic Resonance Imaging



Dobutamine stress magnetic resonance imaging demonstrating abnormal myocardial perfusion in the basal to mid-anteroseptum **(B, C)** and inferolateral walls **(A to C)** as well as the apical lateral wall **(D)**. Findings were consistent with myocardial ischemia in the absence of corresponding delayed gadolinium enhancement.

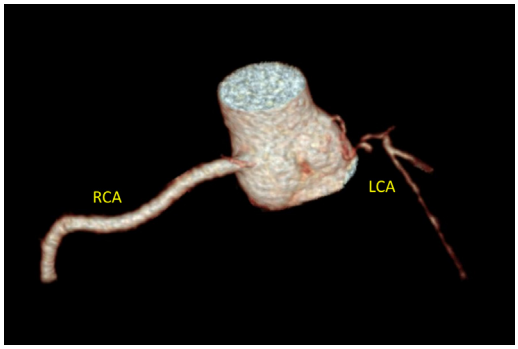
FIGURE 5 Coronary Angiography



(A) Invasive angiogram demonstrating a dominant right coronary artery originating from the left coronary cusp. **(B)** A long myocardial bridge was present in the mid-left anterior descending artery. The instantaneous wave-free ratio of this lesion was 0.86, consistent with ischemia, and normalized on pullback.

and may be preferred if radiation exposure is a significant concern.[4] Stress CMR is particularly useful because of its ability to detect prior infarcts with late gadolinium enhancement.[1] However, it is important to acknowledge that pharmacologic stress may not replicate the level of physiological stress present with maximal exercise. In case 1, this principle was evident with the submaximal heart rate during stress CMR. Thus, maximal-effort exercise stress testing to provoke symptoms and ascertain exercise ECG should be included in the clinical evaluation. However, a single test in isolation is insufficient for risk stratification because a normal exercise stress ECG also does not rule out high-risk anatomy.[4] All clinical risk stratification tests must be integrated with the clinical history. There also remains a place for coronary angiography in CAA evaluation, although this should be reserved for cases in which invasive ischemia assessment (case 2) or high-resolution anatomical detail (case 3) is necessary to guide management.

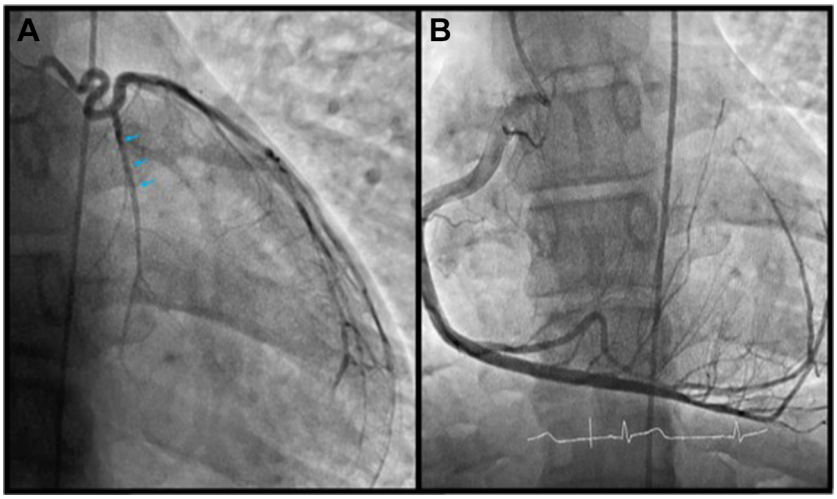
FIGURE 6 Computed Tomography Angiography



3-dimensional reconstruction demonstrating a diminutive hypoplastic left coronary artery. LCA = left coronary artery; RCA = right coronary artery.

For most CAAs, the decision to proceed to surgery and the safety of sports participation comes with clinical uncertainty (excluding anomalous interarterial left coronary artery, which warrants surgical correction[3]). The CAA guidelines are largely consensus and based on epidemiologic SCD studies and autopsy series.[2,4] Ensuring an athlete-centered outcome with SDM is therefore essential in the determination of clinical management, sports eligibility, and clinical surveillance strategies. SDM should include relevant stakeholders who oversee the athlete to ensure that appropriate emergency action planning is present. Although intensive risk stratification augments SDM, the final clinical determination should remain guided by an athlete-centered principle. All parties, including the guiding clinician, must be able to move forward with a management plan that incorporates the athlete's goals and values.

FIGURE 7 Coronary Angiography



(A) Invasive angiogram of the left coronary artery demonstrating a hypoplastic left coronary artery. **(B)** Large dominant right coronary artery with robust septal collaterals to the anterior myocardium and left anterior descending artery.

CONCLUSIONS

CAAs present numerous challenges in the care of affected athletes. Although most are benign, malignant CAAs are associated with SCD. The clinical presentation and differential phenotypes of pathologic CAAs can be variable, with uncertain risks associated with exercise. Whereas imaging and functional assessments facilitate risk stratification, clinical uncertainties remain, and management strategies are often constructed without the benefit of outcomes studies. In most cases, individualized SDM should be relied on in the care of athletes with pathologic CAAs.

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KEY WORDS athletes, cardiac CT, coronary artery anomalies, shared decision making, stress testing

APPENDIX For a supplemental video, please see the online version of this paper.