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EDITORIAL COMMENT

How to Work Up a Case of Sudden Cardiac Arrest in a Young Sportsman*

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Recently, it has become progressively clearer that only 1 specific class of coronary artery anomaly, the anomalous origin of a coronary artery from the opposite sinus of Valsalva (ACAOS), is associated with a potentially serious mechanism of ischemia known as intramural (IM) aortic course, in which the anomalous artery course is located inside the aortic wall. The resulting stenosis is dynamic in nature and related to aortic expansion in systole (varying during the cardiac cycle and more obstructive in systole and with tachycardia, increased cardiac output, and hypertension) (1,2). This behavior is present in 100% of cases of ACAOS with a pre-aortic course, as demonstrated only by surgical inspection (an imprecise method) or intravascular ultrasonography (IVUS) (a precise method) but not by echocardiography, catheter angiography, or computed tomography angiography (CTA), because the spatial definition of such imaging is not adequate for identifying the split aortic wall within which the ectopic coronary artery courses. Furthermore, ACAOS with an intramural course may involve the right coronary artery (R-ACAOS-IM) or left coronary artery (L-ACAOS-IM). Of note, the alternative terminologies AAORCA and AAOLCA (used primarily by pediatric cardiologists and surgeons [3]) and the even more generic AAOCA (which includes all anomalies of origin, all courses, and any artery) encompass various course anomalies (e.g., pre-pulmonary; intraseptal or infundibular; retroaortic, retrocardiac, or periapical), creating significant potential for miscommunication. No other type of ACAOS has the same potential as ACAOS-IM to cause significant ischemia.

The paper by De Olivera Nunes et al. (4) in this issue of *JACC: Case Reports* offers an occasion to consider updated rules for this evolving discipline in sports cardiology. The case report describes a 14-year-old boy who experienced sudden cardiac arrest (SCA) during customary exercise without ever having reported any relevant symptoms. He was found to have congenital R-ACAOS-IM.

CAN A CORONARY ANOMALY'S SEVERITY BE QUANTIFIED?

Coronary artery anomalies are typically qualified as such without a sensitive technique to establish the range for quantifying the implied severity of stenosis. Both CTA and IVUS can potentially do that, although CTA achieves diagnostic definition only with bradycardia (ideally, <60 beats/min) and during end diastole; unfortunately, end-systolic imaging is likely to be inaccurate because of the short opportune time and cardiac motion. The most useful CTA image for quantifying stenosis severity is the right anterior oblique projection in tomographic cross-section (the Angelini-Cheong sign [5]) (**Figure 1**). A ratio of more than 2:1 is suspicious for significant stenosis as evaluated at rest and in diastole. Much more refined imaging of the IM course can be obtained by IVUS (9-to-10 times better temporal and spatial definition), which permits continuous imaging at a rate of 30 images/s. IVUS consistently discredits the older notion that the combined systolic expansions of the aorta and pulmonary

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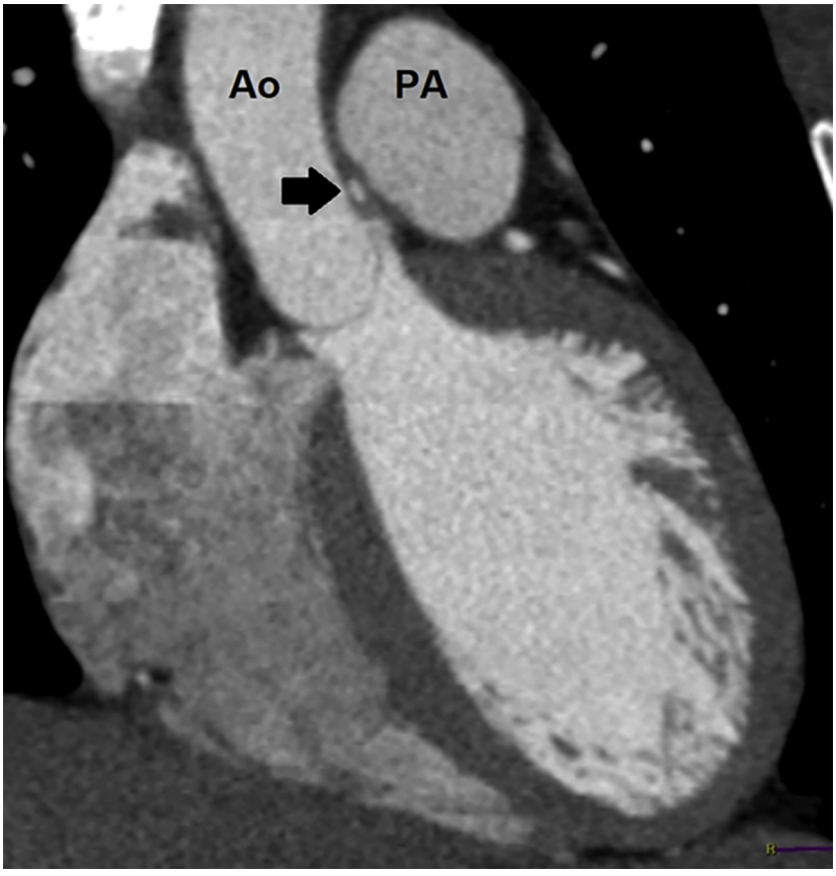
FIGURE 1 Computed Tomography Angiography of ACAOS-IM



A right anterior oblique section at the level of the aortopulmonary sinus shows the proximal R-ACAOS-IM at its narrowest site (the Angelini-Cheong sign) in front of the anterior aortic commissure **(arrow)**. The severity of stenosis can by estimated by computing the ratio between the long and short diameters. In this case, the ratio was 2.4 (moderate severity). Ao = aorta; PA = pulmonary artery; R-ACAOS-IM = anomalous origin of a coronary artery from the opposite sinus of Valsalva (ACAOS) with intramural course involving the right coronary artery.

artery lead to compression of the IM coronary anomaly. On the contrary, systemic pressure in the ectopic artery frequently creates an indentation in the lower pressure pulmonary artery wall.

CLINICAL MANIFESTATIONS OF R-ACAOS-IM

Most young athletes with R-ACAOS-IM perform normally during training and competing and rarely report pertinent symptoms, because a smaller territory is perfused by the right coronary artery than by the left coronary artery. Nonetheless, stenosis exists in every case studied by IVUS and ranges from 10% to 80% at diastole and 20% to 100% at systole in the cross-sectional area. The usual pre-event symptoms include chest pain (which is typical, related to exercise intensity; or atypical, unrelated to exertion), dyspnea, dizziness, and syncope (rare). Acute myocardial infarction also is rare (and never correlated with thrombus formation) (2,3). Sudden cardiac death (SCD) in R-ACAOS-IM (6–8) is exceptional and occurs mostly in males (probably 9 times more frequently than in females) and during exertion (>85% of cases) (6,9).

The case reported by De Olivera Nunes et al. (4) is quite rare, and the accompanying clinical features are important to review. First, no pre-event symptoms were reported (4). Not mentioned is whether the young man was pre-certified for sports participation; in any case, his medical history plus a physical examination (the standard pre-participation screening technique in the United States) usually could not

identify ACAOS, as confirmed by this reviewer's large cardiac magnetic resonance (CMR) imaging-based screening study in students 11 to 18 years of age (10,11). Pre-event symptoms such as those mentioned above are rare before SCD or SCA or syncope but are potential manifestations of R-ACAOS-IM in an exceptionally severe phenotype or exercise conditions.

In cases of SCA, CTA is indicated (New York Heart Association functional class III symptoms) (3,5). The indication for CMR is less certain because its precision is less than ideal and because these individuals have urgent need for final diagnosis. When SCA or syncope has occurred during exertion, especially if accompanied by chest pain, CTA of R-ACAOS is all that is necessary to determine the need for surgery in a young athlete. Additional techniques may be required in adults and in the presence of potentially confounding elements, such as acquired coronary disease or accompanying cardiomyopathy (e.g., in the reported case, arrhythmogenic right ventricular cardiomyopathy might have been suspected). In children and military recruits, IVUS is usually recommended only in the presence of unclear clinical events suggestive but not definitive for ACAOS-IM or of expected exposure to maximal, competitive exertion (2,12). Treadmill exercise electrocardiography or nuclear testing is not usually able to improve diagnosis of severity (2). In a patient with R-ACAOS-IM and ventricular arrhythmia, like that reported by de Oliveira Nunes et al. (4), maximal exertion testing could be life-saving by discovering episodes of ventricular arrhythmia potentially related to right ventricular arrhythmogenic cardiomyopathy.

Reports of the incidence of SCD in R-ACAOS-IM are limited (12). Especially notable are results from 6.3 million military recruits: SCD due to this condition was never reported, whereas 33% of the cases of SCD were due to L-ACAOS-IM in men (no women) (9). In another important study of young athletes (15 to 35 years of age), SCD was 4 times more frequent in L-ACAOS than in R-ACAOS, even though R-ACAOS-IM was found to occur approximately 3 times more frequently than L-ACAOS-IM in a CMR-based population study (10).

Pediatric cardiologists typically do not use IVUS in younger patients, partly due to unfamiliarity with such catheter procedures that are also less useful in children than in adults and because coronary acquired disease is not a factor at a younger age. The additional technical demands of selective catheterization relate to the tangential ostial anatomy that complicates tip catheter selective engagement and alignment. The introduction of dedicated catheters for R-ACAOS-IM cases easily resolved this difficulty (2). In adults, once IVUS has precisely identified the basic anatomical defect (location, length, severity of stenosis in systole and diastole, and ostial involvement), if the indication to treat the defect is well established, stent angioplasty can produce excellent immediate and late results and requires only a few additional minutes (2).

If surgery is performed, it is important to report operative findings, especially those confirming the presence of an IM course and its severity before performing unroofing (although stenosis is difficult to quantify in arrested hearts in the surgical suite (3).

CONCLUSIONS

In the context of preventing serious exercise-related clinical events, the fact that CMR may not be the best approach for quantifying the severity of a patient's R-ACAOS-IM does not exclude it as an excellent technique for screening populations at increased risk, such as athletes. Routine screening CMR has proven reliable for identifying ACAOS-IM without the need for intravenous lines, contrast medium, or ionizing radiation (10). Considering that 0.4% of the general population is born with some form of ACAOS-IM, a potentially high-risk condition that history and physical examination cannot identify, undergoing a CMR-dedicated unit with a competent interpreting team could identify up to 99% of such structural heart conditions and potentially prevent catastrophic surprises (10,11). Given a US population of 330 million, 0.4% represents 1.3 million carriers of ACAOS-IM, a substantial group and not only of concern during their sports years.

In sum, preventing SCD by screening is better than hoping to treat it effectively on the athletic field, where unheralded catastrophes may occur (12).

AUTHOR DISCLOSURES

The author has reported that he has no relationships relevant to the contents of this paper to disclose.

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