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Review

The Trendy Anomalous Aortic Origin of the Coronaries, Should We Start Revising the Pre-Participation Guidelines?

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Abstract: Anomalous aortic origin has become in the last few years the subject of many reports, it is suggested that it is the second most common cause of sudden death among young athletes and school children. Most of patients, will remain asymptomatic until, ischemia is triggered by intense physical activity inducing myocardial hypoperfusion, with subsequent syncope, chest pain or sudden cardiac death. The asymptomatic nature of this anomaly means that even 12-lead electrocardiogram at rest, will miss the diagnosis, which might mandate adding echocardiography or stress-ECG to the guidelines of preparticipation screening.

Keywords: AAOCA; Sudden cardiac death; preparticipation screening

Background

Sudden death in young competitive athletes due to previously unsuspected diseases is a rare but impactful event. The prevalence of athletic field deaths has been estimated at approximately 0.5 per 100,000 per year in high-school-age athletes in the U.S. and 1.6 per 100,000 per year in competitive athletes in Italy [1].

In a study conducted on American youth sports, the overall incidence rate was 1.83 deaths/10 million athlete-years, with the most frequent cause of sudden death being cardiac-related (76%, or 34 out of 45 cases) [2]. In another study by the National Center for the Review and Prevention of child deaths in subjects under 21 years of age, which reviewed 1098 cases, the most common cardiovascular causes of death included congenital heart disease, 40.8%; arrhythmias, 27.1%; cardiomyopathy, 11.8%; myocarditis, 4.6%; congestive heart failure, 3.6%; and coronary artery anomalies, 2.2%. Though the mechanism of sudden death in most cases are believed to be episodic myocardial ischemia, coronary anomalies are amenable to surgical correction, thereby emphasizing the crucial importance of timely identification during life[3].

Coronary anomalies were identified as a contributing factor in 11.8% of fatalities among high school children in a report [4], underscoring the importance of recognizing this issue. The American Heart Association's report [5] indicated that such anomalies account for 19% of deaths in athletes. Angelini and colleagues conducted an angiography study involving 1950 patients to assess coronary

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artery disease, discovering a 5.6% occurrence of coronary anomalies. This indicates that approximately 1 in 20 individuals in the U.S. may have some form of coronary anomalies.

The aim od this perspective/review is to highlight the importance of revisiting the guidelines of preparticipation screening in light of an increasingly recognized cause of sudden cardiac death, namely, the anomalous aortic origin of coronaries (AAOCA) with interarterial course.

Main Body

1-Taxonomy and Nomenclature of Anomalous Origin of the Coronaries

The RCA originates from the aorta's right side through the right aortic cusp, branches into the right marginal and posterior descending arteries, supplying the right atrium, right ventricle, SA and AV nodes, parts of the left ventricle, and posterior septum. The LCA, from the left side arising through left aortic cusp, branches into the left anterior descending (LAD) and left circumflex (LCx) arteries, which supply the left atrium, anterior and lateral left ventricle, and apex[6].

Understanding the intricate anatomy of the coronary arteries allows a greater comprehension of the pathophysiology that leads to sudden cardiac death in this condition. As detailed in the sections above, coronary artery anomalies are changes in the anatomy of the coronary arteries. Coronary anomalies' classification is illustrated in Table 1. [7]

Table 1. Taxonomy of Coronary artery anomalies.

From the aorta
Left main coronary artery arising from the right anterior sinus
Right coronary artery originating from the left coronary sinus
Left circumflex or left anterior descending coronary artery arising from the right coronary sinus
A single coronary artery arising from the right, left and/or non-coronary sinus
Ascending aorta (high aortic origin)
Descending aorta
From the pulmonary artery
Left coronary artery arising from the pulmonary artery (Bland-White-Garland syndrome)
Right coronary artery arising from the pulmonary artery
Left anterior descending coronary artery arising from the pulmonary artery
Both left and right coronary arteries arising from the pulmonary artery
As a branch of another coronary artery
Left coronary artery from the proximal right coronary artery
Left circumflex coronary artery from a right coronary artery
Right coronary artery as a branch of the left circumflex coronary artery
Right coronary artery arising from the left anterior descending artery
From other arteries
Innominate artery
Brachiocephalic trunk
Left mammary artery
Left subclavian artery
Carotid artery
Bronchial artery
From the left ventricle

The most frequently observed pathway for an anomalous right coronary artery (RCA) originating from the sinus of Valsalva is interarterial, which means it runs between the ascending aorta and the pulmonary trunk. This variation is linked to a risk of sudden death in as much as 30%

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of affected individuals. It is thought that during physical exertion, the aorta's expansion can constrict the anomalous opening, leading to decreased blood flow in the coronary artery and increasing the risk of ischemic changes in the heart muscle [8].

In over 75% of patients with the left coronary artery (LCA) arising from the right sinus, the LCA may take an interarterial path, whether as a distinct vessel or as a branch of a common coronary artery. The heightened risk of sudden death is attributed to the sharp angle at the ostium, the "stretching" of the intramural segment, and the compression that occurs between the right and left coronary cusps. Sudden death may happen due to temporary compression of the LCA's anomalous route, which can be triggered by the dilation of the aorta and pulmonary artery during vigorous exercise, leading to torsion or compression of the coronary artery between the aorta and the right ventricular outflow tract[9].

2-Anomalous Aortic Origin of the Coronaries, Relatively Silent Anomalous!

Most cases with anomalous aortic origin of the coronaries are asymptomatic and their first encounter is usually a serious symptom ranging from syncope or ischemic chest pain to aborted or non-aborted sudden cardiac death. The European registry has published its report on this topic recently in 2024. Out of 262 patients, a total of one hundred thirty-nine patients (53.1%) exhibited symptoms, with chest pain being the most prevalent complaint (n = 74, 53.2%), followed by syncope (n = 21, 15.1%). Additionally, seven patients (5%) experienced a myocardial infarction, while two patients (1.4%) had aborted sudden cardiac death. The most observed condition was right anomalous aortic origin of the coronary artery (right-AAOCA), found in 150 cases (57.5%), followed by left anomalous aortic origin (left-AAOCA) in 51 cases (19.5%), and circumflex AAOCA in 20 cases (7.7%)[10].

The American heart association has published a similar article. In half of the cases (n=80, 49%), the diagnosis was found incidentally. The remaining patients exhibited various symptoms: exertional symptoms (n=35, 21%), non-exertional symptoms (n=32, 20%), a family history of cardiovascular disease (n=8, 5%), sudden cardiac arrest or shock (n=5, 3%), and issues related to arrhythmia or bradycardia[11].

This concludes that most of cases with AAOCA will remain asymptomatic until they present with an aborted or sudden cardiac death.

3-Do We Need to Revise the Current Pre-Participation Screening Guidelines?

The current guidelines of the American Heart Association outlines 14 key prescreening elements – such as a history of elevated systemic blood pressure, knowledge of certain cardiac conditions in family members, and the presence of a heart murmur – that are designed to identify, or at least raise the suspicion of, cardiovascular diseases that place certain athletes at risk. Patients would receive ECGs, echocardiograms, and other follow-up tests if the screening indicates the presence of a problem[12,13].

The AHA refuses to implement mass ECG or echocardiography-based screening programs. Implementing a widespread ECG screening program could place a significant burden on the healthcare system, as there may not be enough qualified medical professionals available to accurately interpret the results. It is crucial to highlight that both false positive and false negative results are a concern [14]. A recent systematic review indicated that the false positive rates for physician-interpretated ECGs in athletes typically range from 1.3% to 2.8% but can reach as high as 26.8% among Arab and Black athletes. In contrast, computer-interpretated ECGs show false positive rates between 2.3% and 5%. Additionally, 12-lead ECGs have been associated with a 10% false-negative rate for hypertrophic cardiomyopathy, the leading cause of sudden cardiac death in athletes. Mandating ECGs for all athletes may also be a challenge for children from low-income backgrounds unless there is adequate financial assistance. It is vital to acknowledge this issue, especially since studies indicate that certain racial groups might have a higher risk of sudden death. Therefore, prescreening programs that rely on costly tests must find solutions to address these significant access inequalities.

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In contrast Italy has adopted since 1982 a strict pre-participation program. In the Veneto region of Italy, the occurrence of sudden death (SD) among competitive athletes saw a significant drop of 89% over the 21 years following the introduction of annual screening legislation. The pre-participation guidelines for amateur competitive athletes are outlined by Italian legislation and regional rules. These protocols are conducted by a physician who has obtained a specialty in sports medicine after four years of full-time training, which includes comprehensive training in ECG analysis. The initial assessment comprises a medical history review, a physical exam, a visual acuity evaluation using a Snellen chart, a resting 12-lead ECG, stress testing for ECG, spirometry, and a urine dipstick test [15].

Between the restrictive American guidelines and the rigorous Italian strategy, we still wander and wonder if it is enough.

As mentioned earlier, the second leading cause among young athletes is the anomalous aortic origin of the coronary arteries. This diagnosis remains largely incidental, and it is manifested usually during exercise, which means that a resting ECG might miss the diagnosis[10].

This opens a new debate on the need of adding a stress-ECG or echocardiography to pre-participation screening. This strategy faces debates of cost effectiveness, and also the false positive disqualifications, with the multiple unknowns related to this trendy diagnosis.

Conclusions

Anomalous aortic origin of the coronaries poses itself as an important unknown in the sports medicine field nowadays. It is surrounded and inducing many unknowns:

-How it induces myocardial ischemia, is it coronary compression, or associations with coronary stenosis, or other unleashed mechanisms

-Does it really mandate surgical correction?

-Does it mandate disqualification from sport?

And more importantly, does it imply changing the current guidelines, by adding echocardiography or stress-ECG to the pre-participation screening or not.

Answers to the latter questions can be guided by cost-effectiveness or by the urge not to lose any life during sports participation; and unfortunately, a consensus is far from being reached.

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List of abbreviations

AAOCA	Anomalous aortic origin of the coronaries
AHA	American Heart Association
ECG	Electrocardiogram
LAD	Left anterior descending

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LCx	Left Circumflex
R/L CA	Right/Left Coronary artery
SD/SCD	Sudden death/Sudden Cardiac Death
US	United States

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