EDITORIAL COMMENT

Congenital Coronary Artery Anomalies in Young Patients

New Perspectives for Timely Identification*

Antonio Pelliccia, MD

Rome, Italy

Sudden and unexpected cardiac deaths in young subjects are rare, but tragic events in competitive athletes assume high public profile, and that unavoidably raise questions regarding the underlying pathological causes and clinical strategies to prevent these catastrophes (1,2).

Various pathological investigations have reported that several structural cardiovascular abnormalities are responsible for sudden death in young athletes, but hypertrophic cardiomyopathy is the most common disease in U.S. autopsy-based series, and right ventricular cardiomyopathy predominates in young Italian athletes (1,3–8). Nevertheless, congenital coronary artery anomalies (CCAA) are frequently found (in about 20% of cases), and they represent the second most frequent disease responsible for athletic field deaths (9–14). Specifically, the wrong sinus coronary artery origin, that is, the left main artery arising from the right anterior sinus (ALMCA), and the right coronary artery originating from the left sinus (ARCA), with a proximal course between aorta and pulmonary trunk, are the most frequent occurrences of CCAA.

See page 593

The article by Davis et al. (15) in this issue of the Journal describes the results of prospective echocardiographic identification of wrong sinus coronary anomalies in a large population of children and adolescents. Their contribution is remarkable, and it is likely to generate new clinical interest regarding these malformations, with special attention to the issues set forth below.

Prevalence in the general population. Prevalence of CCAA and, specifically, of the wrong aortic sinus coronary anomalies in the general population is still uncertain. Previous studies suggested that these malformations occur in 0.2% to 1% of a large group of patients undergoing cardiac catheterization (16). Davis et al. (15) in the present article report a prevalence of 0.17%, derived from a population of 2,388 children and adolescents prospectively examined by echocardiography. However, their study population comprised asymptomatic children and adolescents referred for cardiovascular investigation, and thus cannot be considered a true expression of the “normal” population. The prevalence of wrong sinus coronary malformations in a large and unselected young population should likely be less than 0.2%. This hypothesis is also supported by results of our previous investigation: in a population of 1,360 asymptomatic competitive athletes, routinely examined by echocardiography, we found no anomalies of origin and course of major coronary arteries, which is consistent with a prevalence of less than 0.1% (17).

Clinical identification. Wrong sinus coronary anomalies are rarely diagnosed or even suspected during life; this is because of the scarcity of symptoms and inadequacy of routine diagnostic testing (8, 18).

Premonitory symptoms, such as chest discomfort, atypical chest pain, and/or syncope, are reported in less than 30% of patients (9–14,19). Symptoms are more likely to be reported by subjects with ALMCA, and they usually occur in association with exercise, but rarely raise clinical suspicion for CCAA (12,13). Indeed, when young individuals presenting with symptoms are evaluated with 12-lead resting and exercise electrocardiography, ischemic changes suggestive for CCAA are rarely detected. In a recent investigation describing the anatomical and clinical profiles of young athletes with wrong sinus origin of coronary arteries, Basso et al. (19) reported that all the resting 12-lead and exercise electrocardiograms (ECGs) available in the 12 subjects examined during life were within normal limits. Indeed, a review of the literature encompassing the results of exercise electrocardiograms in 18 young patients (<35 years) with CCAA reveals that ischemic changes were present in only four subjects (i.e., 22%), including two who were already symptomatic (19).

Echocardiography, instead, has the potential to address correct diagnosis, because it provides good anatomic definition of the ostium and proximal epicardial course of coronary arteries. In 1,360 young athletes prospectively evaluated by echocardiography, we were able to visualize the ostium and proximal epicardial course of LMCA in 97% and RCA in 80% of subjects (17). Indeed, in young individuals presenting with symptoms or normal 12-lead ECG, echocardiography provided correct identification of wrong sinus origin of the coronary arteries, which was subsequently confirmed by coronary angiography (20–22). Therefore, the present article by David et al. (15) is outstanding because it demonstrates that prospective echocardiographic assessment is an efficient means to detect during life wrong sinus coronary malformations in a large population of children and adolescents. The present study emphasizes the diagnostic role of echocardiography, and it suggests that investigation of ostium and proximal course of coronary arteries should be a routine part of any echocar-

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From the Institute of Sports Science, Department of Medicine, Italian National Olympic Committee, Rome, Italy.
Mechanism(s) precipitating sudden death. The mechanism(s) leading to sudden death in patients with CCAA are triggered by myocardial ischemia. Occurrence of ischemia is suggested by pathological evidence of acute myocardial damage (small infarcts) and/or chronic injuries (patchy areas of myocardial fibrosis) in the area supplied by the anomalous coronary artery (10–12,19). Ischemia is the consequence of anatomical malformations, including the acute angle takeoff of the anomalous vessel, with a narrowed slitlike orifice that collapses in a valvelike manner, thereby limiting the blood flow. Other anatomical features responsible for ischemia are the proximal intramural course of the anomalous vessel, which is squeezed within the aortic wall, and the compression of the anomalous vessel along its course between the aorta and the pulmonary artery, particularly during exercise. However, in a pathology analysis that comparatively assessed the anatomical features of ALMCA or ARCA in 12 patients who died by these anomalies and in 18 patients who died of unrelated causes, Taylor and colleagues (13) found that neither the size of the ostium, the degree of angle takeoff, the length of intramural aortic course, nor the presence of an ostial ridge was predictive for incidence of sudden death.

Whatever the anatomic malformation, patients who died suddenly during exercise have usually done the same amount of exercise or even more strenuous exercise multiple times, without symptoms or physical impairment. It has been suggested, therefore, that ischemia may be caused by sporadic spasm of the anomalous coronary artery induced by endothelial injury (14). Another mechanism may be the occurrence of ventricular tachyarrhythmia, on the basis of an electrically unstable myocardium. In a few patients being monitored with ECG at the time of their sudden death, ventricular fibrillation was the final event (10,14).

Clinical management. Timely identification of patients with wrong sinus coronary anomalies also raises the question of clinical management. Though it is clear that not all individuals with such anomalies are at risk of sudden cardiac death and that many patients have lived a full life and died of unrelated causes, it is also well known that these malformations are among the most common reasons for sudden and unexpected death in young individuals, particularly during sporting activities (3–5,7–14). The paucity of reports describing the clinical profile and efficacy of surgical approach in patients with incidentally discovered CCAA makes the present recommendations for clinical management still controversial. For simplicity, we may consider the most common instances:

1. A young patient (<35 years) with unequivocal diagnosis of wrong sinus coronary malformation, presenting with symptoms and/or showing evidence of myocardial ischemia: In this case, the surgical approach to repair this malformation is mandatory.

2. A young subject (<35 years) with incidental diagnosis of wrong sinus coronary anomaly, in the absence of symptoms or signs suggestive of myocardial ischemia: In this case there is uncertainty regarding the most appropriate therapeutic choice. In making this decision, we have to consider that precise mechanism(s) leading to ischemia and sudden death are still unknown and unpredictable, and these patients cannot be reliably stratified with regard to risk. Therefore, it seems reasonable that these young subjects should not be exposed to the risk of sudden death, and surgical repair of the coronary malformation should be considered. Attention should be paid to the level of the patient’s physical activity, since the greatest incidence of sudden death occurs during heavy physical exertion. Changing to a completely and chronic sedentary lifestyle in children and adolescents is not a realistic option, and the desire to participate in regular heavy exercise or engage in competitive athletics represents a further indication for surgical repair. Heavy exercise programs and competitive sports should be clearly discouraged in young patients with CCAA, at least until surgical correction is performed (24).

3. An adult or older patient with incidental diagnosis of wrong sinus coronary malformations, in the absence of symptoms and inducible ischemia: In consideration that sudden death occurs at a young age, this occasional finding has likely no clinical significance and most probably needs no surgical therapy. However, because the magnitude of the risk remains unknown, individualization of the therapeutic choice is appropriate.

Finally, this author believes that it is now timely and appropriate to establish an international registry of prospectively identified young patients with CCAA in order to achieve a better understanding of the clinical profile and the impact of surgical correction on the natural history of these malformations.

Reprint requests and correspondence to: Dr. Antonio Pelliccia, MD, Institute of Sports Science, Department of Medicine, Via dei Campi Sportivi, 46, 00197 Rome, Italy. E-mail: ant.pelliccia@libero.it.
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