Sudden cardiac death due to myocardial ischemia has been well documented in patients with anomalous left or right coronary arteries arising from the opposite coronary sinus (ACA). Many such patients had been asymptomatic before the fatal event, with no previous diagnosis of coronary abnormalities. Thus, when patients are recognized with this abnormality, aggressive surgical management is often recommended.

The etiology of myocardial ischemia in ACA has been attributed to a number of factors, either singly or interrelated. These include: coronary artery coursing between the aorta and pulmonary artery, slit-like coronary orifice, acute angle of take-off of the anomalous coronary artery from the aorta, and the presence and length of intramural segments of the coronary vessel. A number of studies have been carried out attempting to isolate a key primary factor, but no single component of the anatomy of the coronary artery has been recognized as the definitive cause of ischemia. A slit-like orifice and a narrow intramural distal artery might be most important, but absolute evidence has not been put forward (1).

An anomalous coronary artery from the opposite coronary sinus can be either a left coronary artery from the right (ALCA) or a right coronary artery from the left sinus (ARCA). Patients who have symptoms of ischemia or arrhythmia with either form of the anomaly are candidates for urgent surgical repair. However, decisions regarding the management of asymptomatic patients with these coronary anomalies are less well defined. Significant differences in presentation and clinical course have been noted between patients with ALCA and those with ARCA. The vast majority of sudden deaths have occurred in ALCA patients, whether previously diagnosed or not; sudden death in patients who had been asymptomatic also is far more frequent (2). On this basis, surgical repair upon diagnosis is justified in all ALCA patients. However, operative decisions regarding patients with ARCA who are asymptomatic must be evaluated subject to a risk-benefit analysis relating the occurrence of sudden death to the number of asymptomatic patients who have the anomaly. The operative risks related to ARCA surgery as well as surgical outcome are important considerations in the process.

A right coronary artery (RCA) arising from the left coronary sinus is far more common than the left coronary artery (LCA) from the right sinus. Yamanaka and Hobbs (3) reported 126,595 patients undergoing coronary arteriography over a 28-year period. Of these, 13,686 (1.3%) patients had coronary anomalies noted. One hundred thirty-six (0.17%) had RCA from the LCA sinus and 22 (0.047%) had LCA from the RCA sinus. This indicates a prevalence of ARCA 6 times the frequency of ALCA. These findings are consistent with the unexpected discovery of an RCA from a left coronary sinus on an echocardiogram carried out in a child with an unrelated innocent murmur. This seems to be occurring with increasing frequency in the modern era of constantly improving echocardiographic technology. In contrast, a left coronary from the right coronary sinus is rarely found by serendipity.

The clinical outcome for ARCA is most often benign. The incidence of sudden death in asymptomatic patients with ARCA is extremely low. Over the course of the past 25 years, clinical reports describe 10 patients who died suddenly with RCA from the LCA sinus who were not known to be symptomatic. None were children under 10 years or adults over 30 years of age. In an important study, Eckart et al. (4) screened 6,300,000 military recruits for sudden death over a 25-year period from the Department of Defense Recruit Mortality Registry. There were 126 nontraumatic deaths, of which 86% were related to exercise. In 21, the autopsy findings were that of an LCA from an RCA sinus. No patient had an RCA from a left coronary sinus.

Because most patients who have died with a coronary artery from the opposite sinus were exercising during the fatal event, it is reasonable to specifically assess the risks for ARCA in athletes. Basso et al. (5) reviewed 2 large registries in the U.S. and Italy reporting sudden cardiovascular death in young competitive athletes over a 10- to 20-year period. Twenty-seven deaths were recorded in patients who had no history of symptoms; 14 patients had an LCA arising from the right coronary sinus, and 1 patient had an RCA from the left coronary sinus.

Management of the asymptomatic patient with ARCA creates a classic risk-benefit dilemma: the question of intervention for a common condition that is almost always benign, but with a small risk of sudden death. Are the risks for surgery in a large number of patients who would never need intervention worth undertaking to possibly prevent a
sudden fatal event in a few? Technical success for surgical unroofing of the coronary artery is well reported, and the risk of mortality seems to be low. However, short- and long-term morbidity after surgical intervention have not been hitherto investigated, and such observations would be critical in the decision-making process as to whether to intervene in an asymptomatic child or adult with an RCA from the left sinus. In the present issue of the Journal, Brothers et al. (6) report significant subclinical changes indicative of myocardial ischemia in post-operative patients after surgical unroofing procedures. Nine of 24 patients met criteria for ischemia at a mean of 15 months after surgical repair. These included 8 of 16 patients with ARCA and 1 of 10 with ALCA. Although these findings are of concern, patients with ARCA who have well-defined symptoms and/or laboratory studies indicating ischemia and virtually all ALCA patients will nevertheless require surgical repair, with careful follow-up for late ischemic myocardial changes. However, for the asymptomatic patient with ARCA, this post-surgical report adds credence to a more conservative approach.

It is reasonable to conclude, on a risk–benefit basis, that a patient with ARCA who is asymptomatic with no evidence of ischemia or arrhythmias during stress exercise studies need not undergo elective surgery. If the incidence of 0.17% reported by Yamanaka and Hobbs (3) is indeed a reflection of the general population, there would be 510,000 Americans with this anomaly. If this finding is 10 times the actual value, there still would be 51,000 undiagnosed individuals with RCA from the LCA sinus. This must be considered in the additional context of: 1) 10 sudden deaths in asymptomatic ARCA patients reported in published medical studies over the past 25 years; 2) no reported deaths reported in 6,300,000 military recruits; 3) 1 sudden death documented in 2 large registries monitoring sudden deaths in athletes over a 25-year period; and 4) the present report by Brothers et al. (6) indicating that late myocardial ischemia often occurs after surgical unroofing of the more common and less lethal ARCA.

When an asymptomatic patient with RCA from the LCA sinus who has undergone a full evaluation to be followed without surgical intervention, the patient and his/her family might well be burdened with chronic anxiety, related to the awareness that a sudden cardiac event is possible. Thus, especially in the era of the Internet, it is an especially important challenge for the cardiologist to provide accurate information and perspective for families regarding the risks versus benefit options. Cardiac surgery is not the treatment for anxiety (either on the part of the patient or the physician). Furthermore, intervention should not be recommended on the basis of “defensive medicine.”

If a decision is made to not operate on an asymptomatic, fully evaluated patient with ARCA, the question of exercise restriction must be considered. At present, recommendations regarding exercise restriction as published in the American Heart Association/American College of Cardiology guidelines do not differentiate between ARCA and ALCA. These abnormalities are discussed together under the heading of Anomalous Coronary Arteries from the Contralateral Coronary Sinus (7). The profound difference in risk between the 2 sub-entities is not addressed. Thus, it is likely that, for a vast majority of asymptomatic patients with ARCA, excessive exercise limitations might be imposed, in many cases resulting in quality of life being unnecessarily affected. It would seem reasonable to revise recommendations regarding exercise in patients with coronary anomalies to take into consideration the differences between the LCA from the RCA sinus, which virtually always requires surgical repair, and the more prevalent RCA from the LCA sinus, which is most often a benign abnormality that can be followed without intervention.

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