Surgical Unroofing of Anomalous Aortic Origin of a Coronary Artery: A Single-Center Experience

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Background. Anomalous aortic origin of a coronary artery (AAOCA) has been associated with myocardial ischemia and sudden death. The optimal management of patients with AAOCA is controversial. We examined our experience with surgical unroofing of AAOCA to determine the midterm effect of surgical repair.

Methods. From October 1992 through December 2011, 75 patients with AAOCA underwent surgical unroofing.

Results. Mean age was 39.6 ± 19.6 years; 23 patients (32%) were aged younger than 30 years. Angina, shortness of breath, or syncope was present in 55 patients (72%); 2 (3%) had history of sudden cardiac arrest. Of 40 patients (53%) who had preoperative stress tests, results were abnormal in 20 (50%). Coronary or computed tomography angiography demonstrated an anomalous right coronary artery (RCA) arising from the left sinus in 69 patients (92%) and the left main coronary artery arising from the right sinus in 6 (8%). Two patents (3%) were referred for recurrent anginal symptoms after previous RCA bypass with the right internal mammary artery. Minimally invasive partial upper sternal split was performed in 17 patients (22%). Two patients (3%) needed right internal mammary artery-to-RCA grafting due to flow acceleration at the RCA ostium. There were no early deaths. One late death (1%) occurred related to noncardiac causes. At follow-up (mean, 18 months; maximum, 7 years), all patients remained free of cardiac symptoms.

Conclusions. Surgical unroofing of AAOCA is associated with low morbidity and mortality. At intermediate follow-up, resolution of symptoms and freedom from sudden death can be expected. The threshold for offering intervention should be low.


Congenital anomalies of the coronary arteries have become increasingly recognized with wider use of improved imaging technologies [1, 2]. Although most cases are clinically silent, anomalous aortic origin of a coronary artery (AAOCA) has been associated with an increased risk of myocardial ischemia and infarction, congestive heart failure, and sudden cardiac death (SCD) [3, 4]. In the United States, congenital coronary malformations are the second leading cause of SCD in the young, exceeded only by hypertrophic cardiomyopathy [5, 6]. The estimated incidence of the left coronary artery (LCA) arising from the right sinus of Valsalva (ALCA) is 0.03% to 0.05% and that of the right coronary artery (RCA) arising from the left sinus of Valsalva (ARCA) is 0.05% to 0.1% [7, 8].

Within the multiple anatomic variants of AAOCA, the presence or absence of an intramural or interarterial course may predispose patients to different risks. Given the rarity of AAOCA, that the relative risk of each of these anatomic variants has not been established is not surprising. A complicating issue is that no unifying mechanism has been determined that will predict symptoms or the catastrophic complication of SCD [3, 4, 9]. The presence of premonitory symptoms and exercise stress testing are all inadequate in stratifying SCD risk [9, 10]. Hence, strategies to manage the treatment of patients with AAOCA are not well defined and vary amongst institutions.

Surgical interventions in patients with AAOCA are generally performed to prevent ischemia or SCD, or both. Surgical techniques include coronary artery bypass grafting (CABG), with or without native vessel ligation [11], reimplantation into the correct sinus [12], pulmonary artery translocation [13], proximal coronary artery patch enlargement [14], and unroofing [15]. Coronary unroofing has emerged as the procedure of choice for this congenital anomaly. We examined our experience with surgical unroofing of AAOCA and determined the midterm effect of surgical repair.

Material and Methods

After Institutional Review Board approval, a review of the Mayo Clinic cardiac surgical database identified 75
patients (52 males [70%]), who underwent unroofing for AAOCA between January 1992 and December 2012. The study excluded 12 patients who had associated coronary artery disease in other areas of the coronary distribution requiring simultaneous CABG and patients who had reimplantation or only CABG (9 patients with ALCA). A retrospective review of medical records was performed, collecting demographic information, presenting signs and symptoms, results of diagnostic workup, including stress testing, specific details of the surgical procedure, and date of last follow-up. We defined “cardiovascular symptoms” as the presence of any of the following: exertional chest pain, shortness of breath, syncope, myocardial infarction in the distribution of the anomalous coronary artery, or aborted SCD.

Operative Technique

Our operative technique for surgical unroofing has been previously described [15]. Recently, we have used a minimally invasive partial upper sternal split in these patients (17 of 76 [22%]). A transverse aortotomy was made to gain access to the anomalous intramural portion of the AAOCA. A probe was placed into the coronary artery, and the intraaortic roof of the AAOCA was sharply opened throughout the intramural pathway. The edges were tacked down with fine suture (Fig 1). If the intramural pathway passed just under the superior aspect of the aortic valve commissure, the commissure was taken down and resuspended (Fig 2). If the intramural pathway was lower and there was concern for potential aortic valve damage, the AAOCA was unroofed only in its true sinus, creating a neoostium while leaving the pathway deep to the valve intact (fenestration technique).

Postoperatively, we recommend low-dose aspirin. At approximately 3 months postoperatively, all patients undergo a stress test to look for signs of ischemia and a computed tomography (CT) scan examining the surgical repair to demonstrate a patent ostium, with no ostial stenosis. If the results of these studies are normal, we allow the patient to participate in sports.

Fig 1. (A) Right coronary arising from left coronary sinus with intramural course. (B) Edges tacked down with fine polypropylene sutures after the slitlike orifice is opened over a probe.

Statistical Analysis

The reported data were obtained by record reviews and patient questionnaires. Continuous variables are expressed as means ± standard deviations or medians (ranges). Vital status for all patients was obtained through use the Social Security Death Index. Kaplan-Meier survival curves were created using SAS 9.1 software (SAS Institute, Cary, NC) and compared using a log-rank test.

Results

Patients were a mean age of 39.6 ± 19.6 years (median, 46 years; range, 13 to 70 years), with 23 (32%) aged less than 30 years. Chest pain, shortness of breath, or syncope were present in 55 patients (72%). The most common presenting symptom was chest pain in 52 (68%), with syncope or near syncopal episodes ranking as the second most common symptom in 15 (20%). Two patients (3%) had a previous acute myocardial infarction, and 2 (3%) had a history of sudden cardiac arrest. Two patients (5%) with ARCA were referred for recurrent chest pain symptoms after right internal mammary artery grafting to the RCA. Associated congenital cardiac defects were present in 4 patients (5%), consisting of tetralogy of Fallot in 2, ventricle septal defect in 1, and coarctation in 1.

Of 40 patients (53%) with preoperative stress tests, results were abnormal in 20 (50%). Preoperative coronary angiography or CT angiography demonstrated ALCA in 6 patients (8%) and ARCA in 69 patients (92%). No patients had associated significant coronary artery disease. The anomalous coronary artery arose from the opposite sinus and traveled between the aorta and pulmonary artery with an intramural course in all patients.

All patients underwent surgical unroofing, with only 2 patients (3%) having limited unroofing or fenestration. Operative records or preoperative imaging describe the ostia of the anomalous coronary artery as “slitlike” in all patients. Three patients (4%) had repeat sternotomies, and 10 patients (13%) had associated procedures, including aortic valve replacement for aortic valve
Surgical unroofing of anomalous coronary arteries (AAOCA) has received attention because of association with sudden death, especially in young adults. Anomalous coronary ostia are a recognized cause of sudden death, especially associated with high-intensity exercise. Theories for the pathophysiology associated with this sudden death include an intramural course, the presence of siltlike ostia, marked artery angulation, compression from the pulmonary artery, arterial spasm, and arrhythmia secondary to minor ischemic insults. Improvements in noninvasive diagnostic techniques, such as transthoracic echocardiography and CT angiography, have increased the ability to easily and safely screen for the condition, leading to increased rates of diagnosis.

This study summarizes our experience with 75 patients who underwent surgical unroofing of AAOCA. There were no operative deaths, and no cardiovascular deaths have occurred at a mean follow-up of 1.5 years. Symptoms in 22 patients (29%) were attributable to myocardial ischemia preoperatively by stress test or history of myocardial infarction, without any significant coronary disease, and none of these patients have had a recurrence of their symptoms during the postoperative surveillance period. Most patients have resumed a normal, unrestricted lifestyle. These results indicate that surgical repair of AAOCA is safe and successful in eliminating symptoms of myocardial ischemia.

AAOCA has received attention because of association with SCD in otherwise healthy and mostly asymptomatic individuals. Although 72% of patients in our series had some symptoms, only 50% of patients who had preoperative stress tests had anginal symptoms that were reproducible by the stress test. Electrocardiogram and exercise stress testing are generally unremarkable in patients with AAOCA. Gadolinium-enhanced magnetic resonance angiogram and myocardial perfusion scintigraphy have shown increased sensitivity to pick up scar burden or perfusion defects in the area of the anomalous coronary, where the standard stress test is negative.

The true prevalence of this condition is difficult to ascertain. Published data suggest that the rate of anomalous coronary arteries arising from the opposite sinus ranges from 0.1% to 0.3% [2–5]. Estimates on rates of SCD with this condition come almost exclusively from autopsy or retrospective data. On the basis of several autopsy studies [1, 3, 22–24], mortality “rates” have been reported to be between 0% and 50% with ARCA and between 30% and 100% with ALCA.

Several studies have retrospectively estimated the risk of death with this lesion. Maron and colleagues [5] provided a comprehensive analysis of SCDs among competitive athletes in the United States during a 27-year period. The authors report the incidence of sudden cardiovascular mortality at 0.61/100,000 person-years, of which anomalous coronaries account for ~11% or 0.07/100,000 person-years. Brothers and colleagues [25] provided a comprehensive analysis of SCDs among competitive athletes in the United States during a 27-year period. The authors report the incidence of sudden cardiovascular mortality at 0.61/100,000 person-years, of which anomalous coronary ostia account for ~11% or 0.07/100,000 person-years. Other studies have included a combination of patients with normal and abnormal coronary arteries. Shad et al. [26] estimated a rate of SCD of 0.17/100,000 person-years, which is comparable to our findings.
used these same data to conduct a further analysis and determined that the cumulative risk of death during a 20-year period from the age of 15 to 35 (the highest risk period) in patients with AAOCA was 6.3% for ALCA and 0.2% for ARCA. A comprehensive study was conducted by Harmon and colleagues [26] reviewing SCD in National Collegiate Athletic Association student athletes from 2004 to 2008. The risk if a student athlete has an AAOCA was 1/860 person-years (0.12%). However, an estimation of a risk of SCD by a large prospective study has significant barriers due to rarity of lesions, the overwhelming majority of which remain unrecognized. Further limiting a prospective study design is the expected low risk of clinical events. Therefore, the true estimate of risk of SCD with this condition in not available.

Hence, identifying specific attributes associated with those who die of this condition might help with risk stratification and prognosis. Several authors have attempted to identify specific factors that might portend a higher likelihood of SCD [9, 10, 27, 28]. Unfortunately, attempts to identify the patients at greatest risk for fatal and nonfatal cardiac events have been largely unsuccessful. Coronary artery morphology, the presence of premonitory symptoms, and exercise stress testing are inadequate in stratifying SCD risk, and the general consensus among those who have published on this topic is that more information is needed to assign risk [4].

The only generally accepted risk factor for SCD is age younger than 30 years at presentation [4, 10]. Older patients appear to have a lower risk of SCD, although some risk may still exist. Thus, decision making is extremely challenging in asymptomatic adults with ARCA.

Our current practice is to recommend surgical repair in all patients with ALCA. The risk of SCD in these patients has been shown to be high, even in asymptomatic individuals. Patients with ARCA, who are symptomatic or have a positive results on stress testing or imaging reveals an intramural, interarterial segment with a slitlike ori
cice and hence negative stress test results. We believe that the slitlike orifice with acute take off of the coronary artery in these patients and an intramural course can lead to intermittent ischemia during periods of stressful activity. Intravascular ultrasonography in AAOCA has documented coronary hypoplasia and localized systolic lateral compression of the intramural segment of anomalous coronaries that run within the aortic wall [27]. This degree of compression appears to have individual variations that probably explain the unpredictable response to exercise in this patient group and hence negative stress test results. Also, asymptomatic patients who are not willing to undergo lifestyle modification with restrictions on their activities undergo surgical unroofing.

CABG and reimplantation, although done in the early part of our experience, are no longer used for this anomaly. Early graft failure after CABG has been reported in several cases. Indeed, 2 patients in our series had undergone CABG elsewhere before surgical unroofing. The early failure is due to competitive flow from patent native vessels contributing to graft thrombosis [29, 30]. Hence, prophylactic bypass in the setting of AAOCA should be discouraged. Reimplantation requires extensive dissection and mobilization and does not address the intramural component. Reimplantation avoids the risk of traditional CABG but carries its own inherent risks, including stretching and kinking of the artery as well as anastomotic stenosis.

Aortic valve competency after unroofing necessitating commissure reimplantation has been a concern [31, 32]. Our approach to repair an AAOCA that passes low behind the commissure is to unroof the coronary artery on each side of the commissure. The commissural detachment is done only if it is high up near the top of the commissure. We have not encountered late aortic valve regurgitation in our series. Similar results have been shown in other series with the use of this fenestration approach [33, 34].

At approximately 3 months postoperatively, if patients have a negative stress test result and no ostial stenosis on CT scan, we do allow patients to participate in sports. However, subclinical changes suggestive of ischemia, despite a patent neoostium after the procedure, have been reported [35]. Hence, lifelong surveillance and a close follow-up are required.

This study has some limitations. Although patients in our series had no recurring symptoms suggestive of ischemia after surgical unroofing, the mean follow-up of less than 2 years is short. A longer longitudinal follow-up is needed to see whether these patients are predisposed to an increased risk of coronary ostial problems and atherosclerosis. Moreover, the follow-up is not 100% complete.

In conclusion, surgical unroofing of a coronary artery from the wrong sinus carries low morbidity and mortality. Early results are encouraging, but definitive data suggesting a lifelong reduction of risk of sudden death after unroofing requires a detailed and extensive longitudinal follow-up. Although the rarity of AAOCA prohibits definitive predictors of ischemia, infarction, or sudden death, based on the substantial risk of this anomaly, the threshold for offering surgical unroofing should be low.

References
INVITED COMMENTARY

Anomalous origin of the coronary arteries is a condition with an estimated incidence of 0.03% to 0.1%, and a significant incidence of sudden death among the young population second only to hypertrophic cardiomyopathy. The paper by Sharma and colleagues [1] is a unique single-institution study of patients undergoing surgery for anomalous aortic origin of the coronary arteries in which they analyze 75 patients treated over a 20-year period. There were no early deaths and only 1 late death but follow-up was exceptionally short, with a mean of 18 months and the longest, 7 years.

In their study, the researchers describe several techniques utilized in the past but stress that coronary unroofing of the origin of the anomalous vessel with an intraaortic or arterial course provides superior cardiac stability over the observed period. The study is not