Aberrant Coronary Artery Origin From the Aorta

Report of 18 Patients, Review of Literature and Delineation of Natural History and Management

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SUMMARY To clarify the natural history and management of patients with aberrant origin of a coronary artery from the aorta, we reviewed 18 patients whose right (RCA) or left (LCA) coronary artery arose aberrantly and passed between the aorta and right ventricular infundibulum.

Nine patients had aberrant LCA. Three young males died suddenly after exertion, each with proximal focal LCA stenosis. None of the six adults with angina (ages 36–70 years) studied angiographically had proximal LCA stenosis. Unlike the young, “sudden death-prone” patients in whom coronary bypass of proximal stenosis may have prophylactic value, our older patients with aberrant LCA did not have proximal stenosis or sudden death, and therefore LCA bypass for sudden death prophylaxis is not warranted.

Nine patients (ages 18–60 years) had aberrant origin and course of the RCA. Seven patients studied angiographically because of angina had no focal proximal RCA stenosis, but two patients had hypoplastic RCA ostia. Although a potential concern in these latter patients, sudden death has not been reported with aberrant RCA, therefore in the absence of syncope, RCA bypass for sudden death prophylaxis is not indicated.

BECAUSE MANY PATIENTS undergo coronary artery angiography and surgery, it is important to be aware of even the unusual morphologic variants of coronary artery anatomy, particularly those with clinical significance. Recent reports have identified two groups of patients with aberrant coronary artery origin from the aorta in which myocardial perfusion may be subject to compromise because the anomalous coronary artery passes between the aorta and the right ventricular infundibulum (R Vinf). 1–3 These studies have linked sudden exertional death in young males with one of these variants. 1–14 However, as experience with these anomalies increased, it became evident that they are not always associated with sudden death, and living patients with these variants have been reported. 1–7 It is important for both prognosis and management to differentiate those who are at risk for sudden death from those who are not.

In this report we review our experience with patients in whom an aberrant coronary artery passes between the aorta and RVinf, describe their presentation, pathophysiologic findings, management and follow-up, review the literature and propose guidelines for patient management.

Methods

Between 1973 and 1977, 18 patients with aberrant coronary artery origin from the aorta, in whom the left coronary artery (LCA) or the right coronary artery (RCA) passed between the aorta and the RVinf, were evaluated at the Massachusetts General Hospital. Incidence data concerning this entity is not discernible from our study since many of these patients were referred to this hospital after initial diagnosis.

In five patients the aberrant coronary artery anatomy was found at autopsy and in 13 the diagnosis was established by selective coronary artery angiography. The angiographic diagnosis of aberrant LCA with delineation of the exact course of the aberrant artery and its relationship to the aorta and RVinf was often difficult. This required selective angiography in multiple projections, in particular the right anterior oblique and lateral views, as outlined in earlier reports of this entity. 1–2 A catheter positioned in the pulmonary artery was helpful in delineating the position of the pulmonary outflow tract while performing selective LCA angiography.

The pertinent clinical history and all available pathophysiologic data, including electrocardiograms, treadmill stress tests, myocardial imaging and autopsy findings, were reviewed for each patient.

Results

In nine patients (fig. 1A) the LCA arose from the right sinus of Valsalva or the RCA, and passed to the left and posteriorly between the aorta and the RVinf to reach the area of usual left anterior descending (LAD) and left circumflex (LCX) distribution. In nine patients (fig. 1B), the RCA arose from the left sinus of Valsalva or the LCA, and passed to the right and anteriorly, between the aorta and the RVinf, to reach the usual RCA distribution. Fifteen patients were male and three were female (overall age range 1–70 years).

Aberrant Origin and Course of the LCA (fig. 1A)

We divided these patients into two subgroups on the
basis of age, presentation and pathophysiologic findings.

The first group consisted of three male patients, ages 1, 11 and 17 years (mean 13 years), who died suddenly after acute physical exertion. Their aberrant coronary arteries were identified at postmortem examination. The infant died after prolonged crying but was previously well. The 11-year-old patient had severe chest pain 2 weeks before sudden death, and again on the morning of death; he was evaluated by a physician, but no intervention was prescribed. Electrocardiographic documentation of the mechanism of death was not available for these patients. The 17-year-old had been entirely well until the time of his cardiac arrest while playing soccer; ventricular tachycardia and fibrillation were observed during unsuccessful attempts at resuscitation. Pathologic findings in these three patients were similar, and all had severe focal stenosis of the proximal LCA. In the infant and the 11-year-old, the LCA arose obliquely from the right sinus of Valsalva, from a separate ostium which had a narrowed, slit-like orifice, then gave rise to an otherwise normal LAD and LCX. In the infant, the myocardium supplied by the LCA was normal; however, in the 11-year-old, the anterior left ventricular wall showed histologic evidence of early infarction. In the 17-year-old boy, the LCA ostium was widely patent, but there was an isolated 80% atherosclerotic plaque approximately 1 cm long in the region where the LCA passed between the aorta and the RV inf. This narrowing was associated with a large acute anterior myocardial infarction.

The second subgroup of six patients with aberrant origin of the LCA consisted of five males and one female, ranging in age from 36-70 years (mean 51 years). These patients underwent coronary angiography because of angina pectoris, and none had a history of cardiac arrest or significant ventricular arrhythmia. Unlike patients in the younger subgroup, these patients did not have demonstrable proximal compromise of the aberrant LCA. Four had significant atherosclerotic coronary artery narrowing in the extramural coronary circulation distal to the congenital abnormality. Five patients had a relatively small LCA system associated with a large dominant RCA which supplied the apex and part of the anterior left ventricular wall (figs. 2A and B). In four of the latter, the myocardium in the LCA distribution was abnormal on resting, stressed or paced electrocardiography (fig. 3). Two patients also had abnormal thallium-201 distribution after stress myocardial imaging and two had a regional wall motion abnormality on left ventricular angiography. One patient had severe aortic stenosis secondary to a calcified bicuspid aortic valve and severe distal coronary artery disease, but no compromise of the proximal LCA.

Management of the six older patients with aberrant LCA consisted of intensive medical therapy of angina in five, with adequate response in three. In two patients, angina was refractory to medical management and coronary artery bypass grafting to the LAD and LCX was performed with relief of pain (follow-up 1 and 3 years, respectively). One of these patients had evidence of decreased thallium-201 uptake on a preoperative stress test which became normal 6 months after surgery. The patient with aortic stenosis underwent aortic valve replacement and his angina is now managed medically.

Aberrant Origin and Course of the RCA (fig. 1B)

Nine patients (seven males and two females) had aberrant origin and course of the RCA. These patients were ages 18–60 years (mean 44 years) at diagnosis. In six patients (ages 51–60 years) the aberrant RCA was an angiographic finding noted at the time of evaluation for angina pectoris. In one patient the aberrant RCA was identified angiographically at catheterization for rheumatic valve disease, and in two patients the diagnosis was made at autopsy. No patient had a history of ventricular tachyarrhythmia or syncope and none died suddenly. The two autopsied patients died in the hospital. One (age 18 years) died from aspiration pneumonia after trauma. This patient had no history or symptoms of heart disease. The second (age 60
years), who died after selective LCA angiography, had severe coronary atherosclerotic disease with near occlusion of the proximal LCA and a congenitally narrowed, slit-like RCA orifice (fig. 4). This patient's death was attributed to transient catheter occlusion of the stenotic LCA with severe congenital RCA obstruction. The terminal electrocardiographic mechanism monitored during unsuccessful attempts at

FIGURE 2. Right anterior oblique projection (A), and left anterior oblique projection (B), showing selective right coronary artery (RCA) angiogram and site of origin of the anomalous left coronary artery (LCA). Note large RCA which extends around the apex to the anterior left ventricular wall, in contrast to the relatively small LCA. LAD = left anterior descending coronary artery; Cx = left circumflex coronary artery.
resuscitation was electromechanical dissociation.

Six patients with aberrant RCA (including five patients with angina) had significant atherosclerotic narrowing in portions of their coronary circulation distal to the congenitally aberrant origin. No patient with angina had demonstrable proximal RCA obstruction (figs. 5A and B). One patient had no demonstrable cause for his atypical angina, and responded to minimal medical regimen. Angina in the other five patients responded well to medical treat-

ment. One patient had severe rheumatic aortic, mitral and tricuspid valve disease and underwent successful triple valve replacement.

Discussion

Because the occurrence of exertional sudden death in patients with aberrant LCA is well-established,1-14 questions arise concerning the natural history and management of these patients. Awareness of this entity is paramount in making this diagnosis during life. It must be considered in young patients, particularly males, presenting with exertional chest pain or syncope, or in those successfully resuscitated. However, not all patients with these anomalies die suddenly, and this diagnosis is being made with increasing frequency in patients who have already demonstrated significant longevity free of syncope. Thus, awareness of the clinical spectrum and natural history of these anomalies is essential for appropriate patient manage-

ment.

Aberrant Origin and Course of the LCA

As demonstrated by our aberrant LCA patients and reported by others,2, 9 there are two subgroups, both with the same morphological pattern shown in figure 1A which, however, differ markedly in their clinical presentation, age at recognition, and pathophysiologic findings.

Our three young patients with aberrant LCA, and 17 similar patients previously reported,2, 4-9 make up a homogenous subgroup. The mean age of these 20 patients was 16 years (range 1-36 years). All were male, and all died suddenly after physical exertion. Two of the 20 patients reported severe chest pain within days or weeks before death1, 7 and three had syncope.2, 5. 7 The terminal electrocardiographic mechanism in three patients who were monitored during their acute event was ventricular fibrillation.1, 8 Four patients had histologic evidence of acute myocardial infarction, and a fifth had changes suggestive of early infarction in the distribution of the anomalous LCA.1, 2, 7 Focal proximal LCA obstruction was present in our three young patients and mentioned in the remaining.1, 2, 4-7 although the proposed etiology for this proximal obstruction varied. In some patients the obstruction had an anatomic basis, in
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others it was attributed to a physiologic mechanism, and still others may have had elements of both. Two of our patients had a hypoplastic LCA orifice in the right sinus of Valsalva which was functionally narrowed further by the abrupt angle which the LCA made as it passed to the left, between the aorta and RVinf. It has been postulated that these patients die during exertion because of proximal LCA obstruction secondary to kinking at the LCA orifice caused by the dilatation of the aorta and RVinf associated with an increased cardiac output.2 5 7 Our 17-year-old patient differed from the others described. He had a widely patent LCA orifice but had severe atherosclerotic stenosis localized to the segment between the aorta and RVinf. In this patient, focal narrowing may have been caused by chronic LCA compression between the aorta and the RVinf. This has been proposed by some as an additional explanation for LCA obstruction in these patients.6 7 However, significant LCA compression between the aorta and RVinf in the presence of normal right ventricular pressure seems unlikely.10

In most young patients with aberrant LCA, the diagnosis was made at autopsy. In one, it was made by angiography after successful resuscitation from two episodes of ventricular fibrillation.2 Thus, diagnosis of this anomaly in life was possible in approximately 20% of the young patients in whom exertional chest pain, syncope, or ventricular tachyarrhythmia preceded sudden death. Thus, appropriate use of noninvasive studies, including stress electrocardiography and myocardial imaging, are indicated if the diagnosis is suspected in a young patient after exertional chest pain, syncope or significant ventricular arrhythmia. If these studies are abnormal, coronary angiography should then be performed. If an aberrant LCA is demonstrated in these young patients, surgical bypass grafts to the LAD and LCX for sudden death prophylaxis is appropriate. One such patient has been previously reported.2

The second subgroup of older patients with aberrant LCA is, to our knowledge, the only such group of patients in whom an aberrant LCA was identified in life. All were identified by coronary angiography performed for evaluation of angina pectoris. None had a history of syncope, ventricular tachyarrhythmia or cardiac arrest. Twenty-nine similar patients (mean age 56 years) with this anatomy who did not have syncope or sudden death but were identified at autopsy examination have been reported.2 10-14 Not surprisingly, in view of the age of these patients, those with angina had significant coronary artery disease not related to their congenital abnormality.12 14 The follow-up of our older patients is 1–4 years, therefore, definitive statements concerning overall longevity cannot be made yet.

The most obvious differences between patients with aberrant LCA who die suddenly and those who do not are the acute clinical presentation, youth and the presence of proximal anatomical or physiological LCA obstruction in the former group in contrast to the latter. Unlike the young patients, the older patients do not seem to be at risk for sudden death and do not have demonstrable proximal LCA obstruction.5 13-14 Therefore, we feel that management of older patients should be directed at their presenting problem, e.g., angina pectoris, and not specifically at sudden death prophylaxis. In four of our patients, initial medical treatment of angina was successful. However, angina was refractory to medical treatment in two others and necessitated aortocoronary artery bypass surgery to which each responded well.

The etiology for angina in our older patients with aberrant LCA appeared to be related to their abnormal left ventricular myocardium. Although the exact nature of these abnormalities is unclear, ischemia was probably present in some, as demonstrated in one of our patients by ischemic ST-T-wave changes which were inducible at an increased heart rate (fig. 3), and by a thallium-201 stress imaging abnormality which was present preoperatively and disappeared after coronary bypass surgery. A relatively small LCA system was noted in four of our patients and has also been described by others.5 6 10-12 We do not know whether the relatively small size of the LCA is related to the aberrant origin of the LCA, as suggested by its frequent occurrence in this entity, or is merely secondary to the presence of a large, dominant RCA supplying the left circumflex coronary artery territory.

Aberrant Origin of the RCA

Our nine patients with aberrant RCA, combined with 18 previously reported autopsied patients,2 had a mean age of 54 years, and none had been known to have syncope, related ventricular tachyarrhythmia or sudden death. Therefore, it would seem that aberrant RCA usually has a benign clinical course.

Considering the ages of these patients (mean 54 years) it is not surprising that coronary artery disease was common. Because sudden death in patients with anomalous RCA is rare, coronary artery bypass as prophylaxis for sudden death does not seem indicated. Management of these patients should be directed at controlling angina, when present, which we accomplished successfully with standard medical regimens in all of our patients. However, as shown by our two patients with hypoplasia of the RCA orifice (fig. 4), proximal RCA stenosis may be severe and could have dire consequences, particularly if associated with a dominant RCA.

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FIGURE 5. Right anterior oblique (A) and left anterior oblique (B) projections showing selective left coronary artery (LCA) angiogram in a patient with anomalous origin of the right coronary artery (RCA). Note absence of proximal RCA compromise (arrows).
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