Aberrant Coronary Artery Origin From the Aorta
Diagnosis and Clinical Significance

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SUMMARY
Twenty-one patients with anomalous coronary artery origin from the aorta are discussed, and the cases reported in the literature are reviewed. The left anterior descending (LAD) and left circumflex (Cx) arteries arose aberrantly from the right sinus of Valsalva of the aorta (RSV) in six patients. In four of these patients the connecting branch from the anomalous origin passed anterior to the aorta and the right ventricular infundibulum (RVinf), and in two patients, this branch passed between the aorta and RVinf. In 11 patients only the Cx was aberrant, and arose either from the RSV directly or from the right coronary artery (RCA), and passed posterior to the aorta and RVinf. In four patients the RCA arose aberrantly — in three from the left sinus of Valsalva of the aorta passing anteriorly, and in one from the left Cx passing posteriorly.

Aberrant coronary artery origin from the aorta had clinical consequences only when the branch connecting the LAD and Cx to the RSV passed between the aorta and RVinf. Both our findings and those reported in the literature associate this variant with exertional sudden death in young persons. Anomalous coronary patterns can be delineated readily by selective coronary cineangiography, and as illustrated, the right anterior oblique projection can readily distinguish those prone to sudden death from the clinically insignificant variants. The former can be corrected with coronary artery bypass surgery.

Additional Indexing Words:
- Coronary artery anomalies
- Sudden death
- Congenital heart disease
- Exertional death
- Coronary artery angiography

As the roles of coronary artery angiography and coronary surgery continue to be defined, identification of variations in the course and origin of the major coronary arteries and knowledge of the clinical significance of these variants become important. Numerous cases have been reported of anomalous origin of the coronary arteries from the aorta, but the rarity of these lesions has obscured both their anatomic and clinical delineation. In the present study we describe the spectrum of aberrant origin of coronary arteries from the aorta seen in 21 patients and discuss the pertinent clinical considerations related to these patterns.

Methods
Cases were obtained from review of the files of the Cardiac Catheterization Laboratory at the Massachusetts General Hospital and from review of the records of the Congenital Heart Disease Research and Training Center at the Hektoen Institute for Medical Research, Chicago, Illinois. Twenty-one cases were identified. In 9 of these the diagnosis was confirmed by autopsy examination and in 12 by selective coronary angiography. The latter patients were referred for coronary angiography because of histories suggestive of congenital coronary heart disease, and therefore are a highly selected population. In all cases past medical records were reviewed and pertinent clinical correlations were made. Cases were divided into three categories: (1) those in which both the circumflex (Cx) and the left anterior descending (LAD) coronary arteries arose aberrantly from the right sinus of Valsalva (RSV) of the aorta; (2) those in which only the Cx arose aberrantly; and (3) those in which only the right coronary artery (RCA) arose aberrantly.

Results
Aberrant Origin of the LAD and Left Cx — 6 Cases
These variants are illustrated in figure 1a and b. In four cases (fig. 1a) the vessel connecting the LAD and Cx to their aberrant origin in the RSV passed anterior to both the aorta and the RVinf. In these patients, selective right coronary angiography in the right anterior oblique (RAO) projection shows this connecting branch passing anteriorly across the RVinf to give rise to the LAD and Cx (fig. 2a). In these patients, the average age was 55 years, three were men, three...
CORONARY ANOMALIES

ABERRANT ORIGIN OF THE LAD and LEFT CX (6 CASES)

ABERRANT ORIGIN OF THE LEFT CX (11 CASES)

ABERRANT RCA (4 CASES)

Figure 1

Diagrammatic representation of aberrant coronary artery patterns. A: aorta; RV inf, right ventricular infundibulum; R, right sinus of Valsalva; L, left sinus of Valsalva; N, noncoronary sinus of Valsalva.

Figure 2

Selective RCA angiogram from a patient with aberrant origin of the LAD and Cx viewed in (a) the right anterior oblique, and (b) the left anterior oblique projection. LCA, connecting branch to LAD and Cx.
Aberrant Origin of the Cx — 11 Cases

These variants are illustrated in figure 1c and d. In these, the left Cx arose aberrantly either from the RSV itself (five cases) or from the RCA (five cases), and the LAD arose directly from the left sinus of Valsalva.

In these patients, the aberrant Cx passed posterior to the aorta traversing the posterior atrioventricular groove to reach its usual distribution (fig. 4a and b). The LAD arose normally from the left sinus of Valsalva; however, a Cx origin from this vessel was conspicuously absent in both oblique projections (fig. 4c and d). The average age of these patients was 54 years, eight were men, five had old myocardial infarction, seven had angina pectoris, and seven had significant coronary stenosis. In these patients, no evident relationship existed between the aberrant coronary pattern and coronary or myocardial lesions.

Aberrant Origin of the RCA — 4 Cases

These variants are illustrated in figure 1e and f. In these, the RCA arose from the left sinus of Valsalva in three (fig. 1e), and in one, there was no RCA, and its usual distribution was supplied by an extension from the left Cx which continued past the posterior descending branch (fig. 1f). In these patients the average age was 53 years, all were men, one had aortic stenosis, one had asympotic septal hypertrophy, two had old myocardial infarction, two had angina pectoris, and two had significant coronary artery disease.

Discussion

Anomalous coronary origin from the pulmonary artery has been well described, as has anomalous coronary origin from the aorta complicating other congenital cardiac anomalies. Aberrant origin of coronary arteries from the aorta in the absence of associated congenital heart disease has also been reported by many; however, the clinical significance of these lesions has received little attention. The incidence of aberrant coronary artery origin from the aorta in our catheterization laboratory population was 0.6%, which approximates the findings of others. Anomalous origin of the Cx alone was most common, followed by anomalous origin of both the Cx and LAD, and then of the RCA alone. These patterns and rare additional variants have all been reported by others. Seventeen of our 21 patients were men, and a male predominance has also been noted by others. Excluding our 11 and 17-year-old patients who died suddenly, the mean age of our patients was 56 years. Our high incidence of coronary and myocardial disease is explained by the selected nature of the catheterization laboratory population.

For clinical purposes, the variations in aberrant left coronary origin may be subdivided according to the course of the aberrant vessel in relation to the aorta and the RVinf. Three primary courses were found. In the first group (fig. 1a), both the LAD and Cx were aberrant, and the branch connecting these vessels to the anomalous origin from the RSV passed anterior to the aorta and RVinf. In the second group (fig. 1c and d), the Cx alone was aberrant, and it passed posterior to the aorta and RVinf. In both of these types, longevity was normal, and the clinical presentation was not related to the aberrant coronary pattern, nor was there an evident relationship between the aberrant vessel and coronary or myocardial disease. Thus, in these patients, the coronary anomaly had no clinical consequences. As discussed by others, however, delineation of the aberrant coronary course and origin is important in those having cardiac surgery to avoid severing a major coronary branch, as well as to avoid exclusion during coronary perfusion of an aberrant Cx arising from the RCA.

In contrast to the above, in our study (fig. 1b) and in those reported in the literature, sudden exertional death occurred in all in whom the branch connecting the Cx and LAD to the aberrant origin in the RSV passed between the aorta and the RVinf. In addition, our two patients and five of the six reported in the literature were adolescent boys, and the sixth was a 26-year-old man. It is thus critical to identify and correct these anomalies when possible. Known prior symptoms were present in one of our patients but were rare in those in the literature. Premortem detection of this anomaly would therefore generally be fortuitous, and we are not aware of any patient in whom this diagnosis was made before death. However,
Figure 4

Selective RCA angiogram from a patient with aberrant origin of the Cx, viewed in (a) the right anterior oblique and (b) the left anterior oblique projection, and selective left coronary artery angiograms viewed in (c) the right anterior oblique and (d) the left anterior oblique projection.

Awareness of the association of this pattern with sudden exertional death, particularly in young males, is important and should be considered in evaluating young patients with acute myocardial infarction, angina pectoris, or cardiac arrest, as it is potentially correctable. Aorto-coronary artery bypass to the Cx and LAD would obviate distal compromise.

When the usual left coronary ostia cannot be located, selective sinus or aortic root angiograms are appropriate to locate the coronary ostia. In the right anterior oblique projection, the relationship of the connecting branch to the aorta and the RVinf is seen clearly, and as diagrammed in figure 5, this relationship can be delineated and the three patterns differentiated. The true lateral projection would also differentiate these patterns.

If the left Cx is not visualized during selective left coronary angiography, aberrant origin either from the RSV or from the RCA should be considered before assuming occlusion of the Cx at its normal origin. In these patients, it is important to verify that the catheter has not passed into the LAD beyond the
origin of the Cx, and care is also needed to avoid confusing a posteriorly placed diagonal branch with the normal Cx (fig. 4c and d).

We were not able to study the physiology involved in our subjects who died suddenly, nor has it been studied in cases previously reported. It is possible that external compression of the aberrant connecting coronary branch which is protected only by adventitia and epicardial fat as it passes between the aorta and RVinf causes obstruction to coronary flow and ischemia. However, in the absence of severe pulmonary hypertension, systolic pulmonary artery pressure should be lower than coronary artery pressure, and diastolic compression would be unlikely.

One experimental study using postmortem perfusion actually failed to show coronary compression despite elevated great vessel pressures. It is possible that with exercise and elevation of both systemic and pulmonary artery pressure, kinking of the aberrant coronary artery occurs as the great arteries dilate secondary to increased cardiac output and force the aberrant coronary artery to turn more acutely to pass between them.

There have been cases reported in which the branch connecting the aberrant origin in the RSV with the LAD passed between the aorta and the RVinf at a lower level where it was imbedded in the musculature of the interventricular septum. These subjects died as adults (age 45–65) and in only one was death attributed to cardiovascular disease. It is possible in these patients that the muscular septum protects the aberrant coronary branch from angulation.

In patients with anomalous origin of the RCA (fig. 1e), as the RCA crosses from its aberrant origin in the left sinus of Valsalva, it also passes between the great vessels and therefore could be subject to compression if this were the mechanism involved. Sudden death, however, has not been reported in these patients, and their average age in our study was 51 years.

Anomalous origin of the coronary arteries from the aorta is an extremely rare cause of sudden death in the general population, and in a recent study of prehospital sudden death, no case was found in 220 autopsies. However, in a selected series of young persons dying suddenly following exertion, a small number of deaths have been attributed to coronary artery anomalies.

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