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Ectopic Origin of a Coronary Artery from the Aorta*

Sudden Death in 3 of 23 Patients

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Ectopic origin of a coronary artery from the aorta is uncommon. In the last decade, its importance as a possible cause of sudden death has been documented. Initially, only the left coronary artery was implicated. Lately, the ectopically arising right coronary artery has also been shown to be involved in cases of sudden death. We reviewed the pathologic anatomy in 23 cases of ectopic origin of a coronary artery from the aorta. In three of these cases, death could be attributed to ectopic origin of a coronary artery. In one case the left coronary artery arose from the right aortic sinus,

Ectopic origin of a coronary artery from the aorta is uncommon, and instances of sudden death have been observed in some cases.^{1,2} The purpose of this communication is to report three additional instances of sudden death. In addition, variations of ectopic origin of a coronary artery from the aorta will be presented.

The location of a coronary arterial ostium was judged to be ectopic when it was located a significant distance laterally or vertically from its normal position. A coronary ostium was considered vertically ectopic if it arose more than 0.5 cm above the sinotubular junction of the aorta. Ostia located just above the aortic sinus were considered variants of the normal and not included. Lateral ectopia was judged to be present if a coronary artery arose from an aortic sinus which did not normally give rise to that artery, above an aortic valvular commissure, or as an anomalous branch of a normally positioned coronary artery.

The pathologic specimens in this study were derived from the Cardiovascular Registry of United Hospital, St. Paul. We were able to identify 28 cases catalogued with a primary or secondary diagnosis of ectopic origin of a coronary artery from the aorta. We did not include any specimen with additional congenital cardiac anomalies or with extensive cystic medial necrosis of the aorta. Also, we did not include cases in which locations

and in two cases the right coronary artery arose from the left aortic sinus. In one of these, a scar of healed myocardial infarction was present in the inferior wall of the left ventricle. The possible mechanism for sudden death in these cases is reviewed and emphasis placed on the theory that the acute angle at which the ectopic artery leaves the aorta results in a flap-like mechanism at the arterial ostium. Ostial stenosis by the flap could be a significant factor in causing myocardial ischemia in some patients and also in sudden death.

of the coronary ostia were normal but the ostium was stenotic or atretic. Of the 28 cases which fulfilled the requirements of this study, 23 specimens were available and constitute the basis for this study.

Our observations will be concerned first with three cases of sudden death. This coverage, in turn, will be followed by a review of the various locations of ectopic origin from the aorta among the 23 cases that constitute the basis for this study.

CASE REPORTS

In each case with sudden death, the subject was white (one female and two male patients).

CASE 1

While playing basketball, a 15-year-old girl was noted suddenly to become pale. She collapsed while walking off the court. Despite attempts of immediate cardiopulmonary resuscitation, she could not be revived and was pronounced dead on arrival at the hospital's emergency room. The patient had not complained of any particular symptoms prior to her collapse. She had no known medical problems and had no history of chest pain or syncope. Previously, she had participated with no ill effects in athletics, including gymnastics, volleyball, and track.

Autopsy revealed that the right coronary artery and the left main coronary artery each arose from the right aortic sinus (Fig 1). The left coronary artery left the aorta in an oblique fashion. This resulted in the formation of a flap of tissue near the arterial ostium (Fig 2). The flap was composed of aortic and coronary arterial tissues. The coronary arteries were devoid of atherosclerosis, and no myocardial lesions were observed. The lungs were edematous.

CASE 2

A 16-year-old boy had a strong family history of premature coronary atherosclerotic disease; however, the subject was considered to be in good health, without significant illness in the past. One day after completing strenuous gymnastic exercises, he sat down and shortly thereafter collapsed. In spite of early attempts at resuscitation, the subject did not respond and was pronounced dead.

The autopsy showed no recognizable coronary atherosclerosis.

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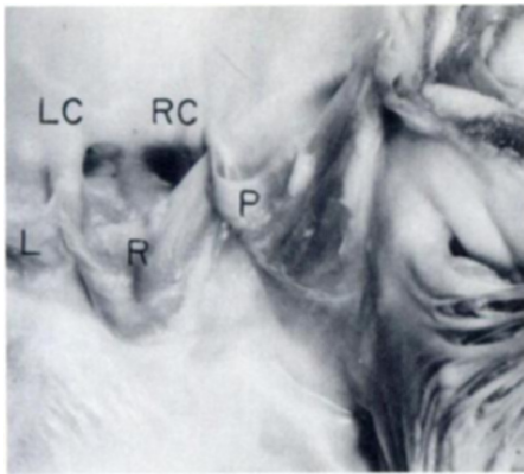


FIGURE 1. View of left ventricular outflow tract, aortic valve, and ascending aorta. Ostia of both left and right coronary arteries lie in the right aortic sinus. L, P, and R, Left, posterior, and right aortic cusps, respectively; LC, left coronary artery; and RC, right coronary artery (case 1).

Both coronary arteries arose from the left aortic sinus. The left coronary artery arose from about the center of the sinus, while the right coronary artery arose from the periphery of the sinus close to the left-right aortic valvular commissure (Fig 3a). From its origin the right coronary artery coursed toward the right, posteriorly to the

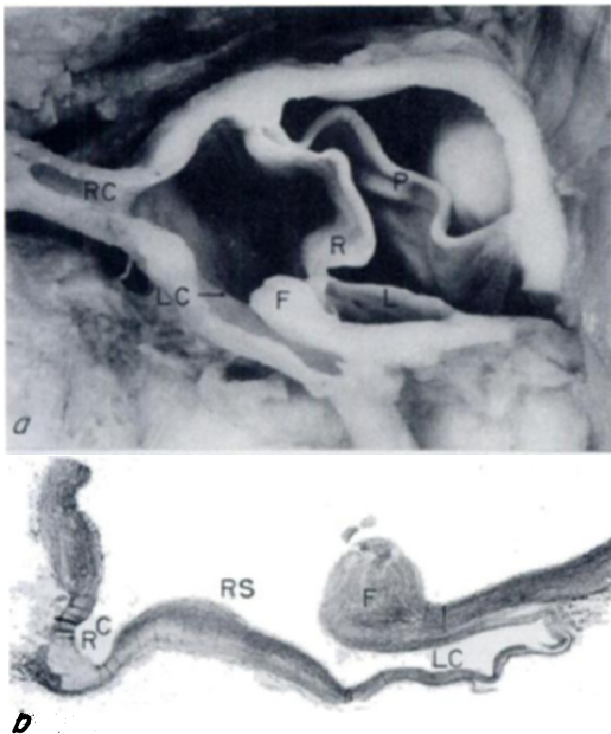


FIGURE 2a (top). Gross view of cross section of ascending aorta at upper levels of aortic valvular insertions. Right and left coronary arteries (RC and LC) each arise from right aortic sinus (RS). As left coronary artery proceeds toward patient's left, it makes oblique angle with aorta and creates flap (F) in relation to its ostium. b (bottom). Section taken through same plane as that shown in Figure 2a to demonstrate wall of right aortic sinus from which arise right and left coronary arteries. Oblique course of ectopically arising left coronary artery creates, along with aortic wall, a flap (elastic tissue stain, original magnification $\times 5$). L, P, and R, left, posterior, and right aortic cusps, respectively (case 1).

pulmonary trunk to reach the right atrioventricular sulcus. The oblique course of the proximal segment of the right coronary artery was responsible for creation of a flap near the arterial origin. The flap was composed of a common structure formed by the adjacent elements of the arterial and aortic walls (Fig 3b). The myocardium showed one abnormality, namely, a scar measuring about 1.2 cm lying in the basal half of the left ventricular inferior wall.

CASE 3

One morning, a 20-year-old man was found dead in bed at home. He had been last seen alive late the night before. The victim had chronic asthma and, when found, had a nebulizer in his hand. There was no evidence of a struggle suggestive of respiratory distress.

At autopsy the lungs were shown to be congested, and there was significant retained mucus in the bronchioles bilaterally. The left and right coronary arteries each arose from the left aortic sinus (Fig 4a). Neither atherosclerosis nor other abnormalities were noted. Toxicologic studies revealed the presence of the nebulizer propellant in the victim's blood. As it left the aorta, the right coronary artery immediately angled to the right, passed between the aorta and the main pulmonary artery, and then followed a normal distribution (Fig 4b).

ANATOMIC TYPES OF ECTOPIC ORIGIN

Of the 23 subjects on which this study is based, 17 were male and six were female subjects. The age at the time of death ranged from 15 to 81 years, with a mean age of 57 years. In each of 18 cases, the cause of death did not appear to be related to the ectopic coronary arterial origin. Two other individuals died following replacement of the aortic valve for stenosis. In each of these, the ectopically arising coronary artery had not been perfused during operation, and acute myocardial

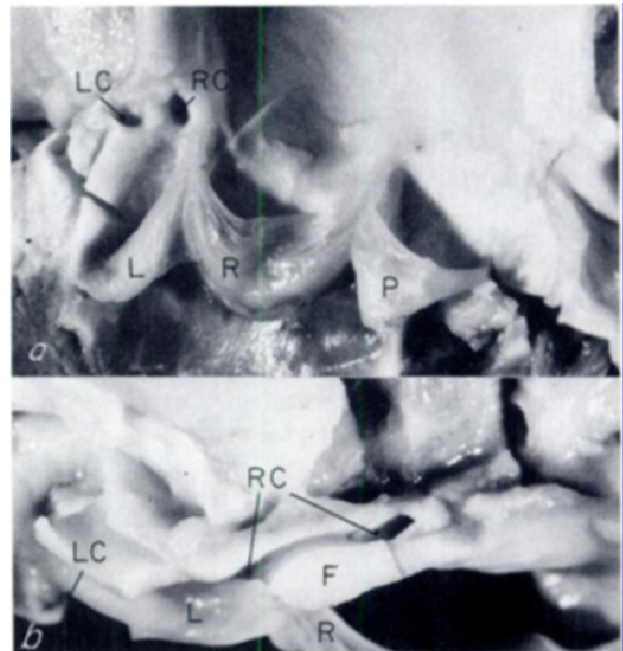


FIGURE 3. Origin of both coronary arteries from left aortic sinus. a (top). Opened aortic origin and valve. b (bottom). Transverse section of segment of aortic wall at upper level of sinuses. Left and right coronary arteries arise from left sinus. Oblique course of proximal segment of right artery is responsible for formation of flap of tissue. F, Flap; L, P, and R, left, posterior, and right aortic cusps, respectively; and LC and RC, left and right coronary arterial ostia, respectively (case 2).

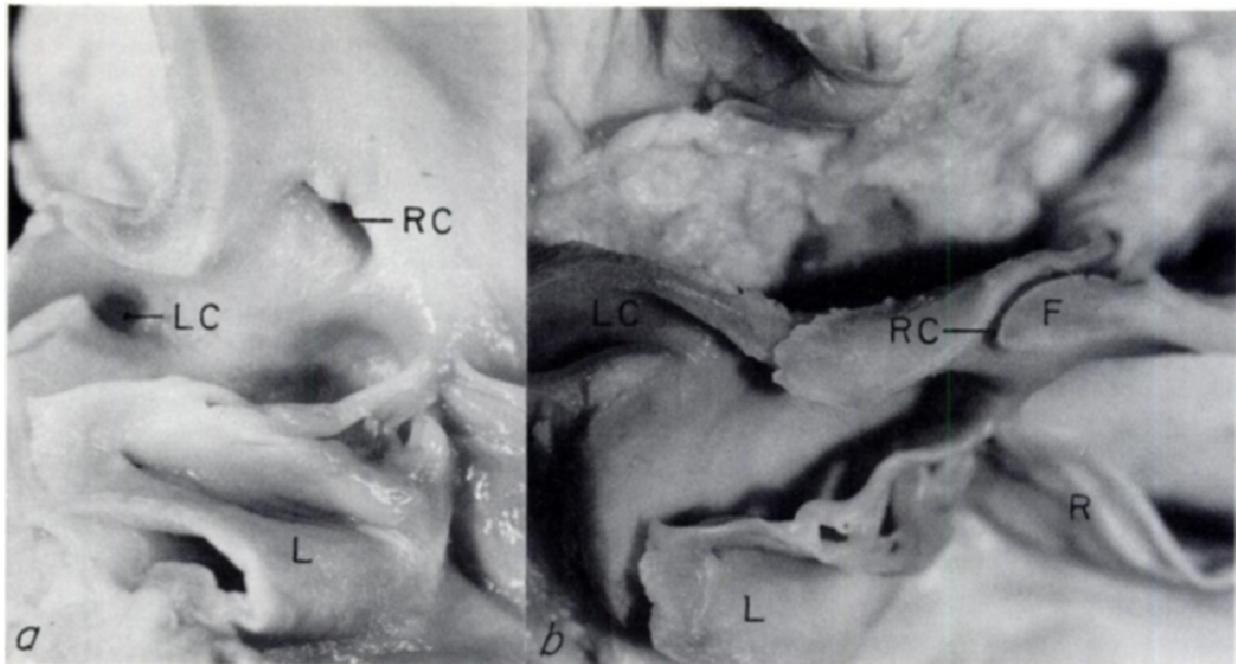


FIGURE 4a (left). Portion of aortic valve and ascending aorta. Left and right coronary arteries (LC and RC) each arise from left aortic sinus. Normal ostium of left coronary artery has been unroofed. Ostium of right coronary artery is slit-like in shape. b (right). Ascending aorta has been sectioned transversely above sinuses to extend into two coronary arteries. As right coronary artery courses from its ectopic origin, it ascribes oblique course through aortic wall, giving rise to flap (F) that contributes to slit-like shape noted in Figure 4a. L and R, left and right aortic cusps, respectively (case 3).

infarction was a complication. The specific details in the three subjects in whom sudden death occurred have been described.

The locations of the ectopic coronary ostia in our 23 specimens could be grouped into nine separate patterns, as shown in Figure 5. Of these cases, 15 demonstrated only lateral ectopia, five involved vertical ectopia, and in three cases the ectopic ostium was displaced both laterally and vertically.

The left coronary system was involved in 12 cases and the right system in 11. The main coronary artery arose from the right aortic sinus in two cases and abnormally high above the left sinus in three. In seven cases the circumflex artery arose either from the right aortic sinus (five cases) or from the proximal right coronary artery (two cases). The right coronary artery arose from the left aortic sinus in five cases, from the posterior sinus in one case, abnormally high above the right sinus in two cases, above the right-posterior commissure in two cases, and above the right-left commissure in one case.

Based on the gross appearance of the ectopically positioned coronary ostia, we observed that in some cases a slit-like orifice was present (Fig 2). In all cases the slit-like configuration of the ostium was a result of the acute angle at which the ectopic artery left the aorta (Fig 2b). Although this type of orifice was seen with several different patterns of ectopically positioned ostia, it was consistently present in distinct lateral displacement, namely, when the left main coronary artery arose from

the right aortic sinus or when the right coronary artery arose from the left sinus. Each of our three cases of sudden death exhibited slit-like ostia.

DISCUSSION

With the advent of cardiac surgery, ectopic origin of a coronary artery from the aorta became recognized as a potential cause of surgical morbidity and mortality. Failure to recognize an ectopic coronary ostium resulted in instances of obstruction of a coronary ostium by a valvular prosthesis or failure to perfuse such an artery during cardiopulmonary bypass.^{3,4}

In addition to reference to iatrogenic surgical complications, the medical literature of the last decade contains multiple case reports of ectopic origin of the coronary artery from the aorta as a cause of sudden death.^{1,2} An excellent review of this subject was written by Cheitlin et al.⁵

Until recently, all cases reported in the literature of sudden death in association with ectopic origin of a coronary artery from the aorta were those in which the left main coronary artery arose from the right aortic sinus⁵ and coursed anteriorly and to the left between the aorta and the pulmonary trunk. The condition of the right coronary artery arising from the left aortic sinus has generally been regarded as a benign variant of normal anatomy.² Roberts et al⁶ reviewed the findings in 26 cases of origin of the right coronary artery from the left sinus reported prior to their study and found no case in which cardiac dysfunction or death

could be attributed to that condition. Roberts and associates⁶ studied their ten post-mortem cases in which the right coronary artery arose from the left aortic sinus. In seven of these, no symptom of cardiac dysfunction was evident; however, each of the other three subjects had died suddenly. The coronary anomaly was the only significant abnormality at necropsy. In two of the three patients, manifestations had been present during life. One had suffered from recurring ventricular tachycardia, and the other one had had typical angina pectoris. In the third case of sudden death reported by Roberts et al,⁶ this condition was the initial manifestation of cardiac dysfunction.

Roberts and associates⁶ also analyzed published data in 31 cases in which the origin of the right coronary artery from the left aortic sinus was detected by coronary angiography. Among these cases, they found nine that

displayed symptoms of cardiac dysfunction while significant coronary atherosclerosis was absent. Bloomfield et al⁷ described a patient with severe angina pectoris who was free of significant atherosclerosis. They attributed the symptoms to compression of the right coronary artery as it coursed between the aorta and right ventricular infundibulum after arising from the left coronary artery. This supposition was based on angiographic evidence of compression of the vessel. Keren et al⁸ also demonstrated cyclic systolic compression of an aberrant right coronary artery that was "squeezed" as it coursed between the great arteries during systole. Liberthson et al⁹ reported the findings in an infant whose right coronary artery arose from the left aortic sinus and who died suddenly with acute myocardial infarction. Hanzlick and Stivers¹⁰ described sudden death in a marathon runner. All cardiac

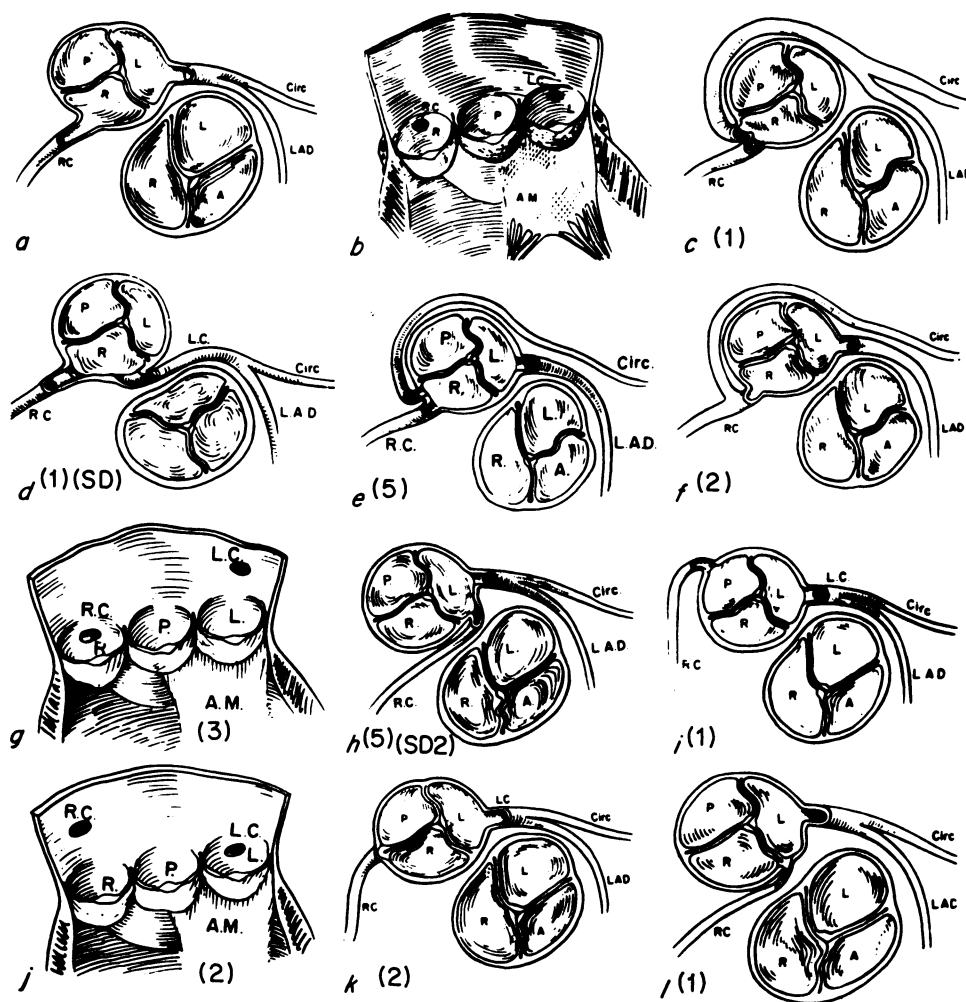


FIGURE 5. Patterns of ectopic origin of coronary arteries from aorta (A) in 23 cases. a and b, Normal. c, Left coronary artery (LC) from right aortic sinus (R) (posterior course) (one case). d, Left coronary artery from right aortic sinus (anterior course) (one case) (sudden death [SD]). e, Circumflex coronary artery from right aortic sinus (five cases). f, Circumflex coronary artery from right aortic sinus (two cases). g, Left coronary artery from above left aortic sinus (L) (three cases). h, Right coronary artery from left aortic sinus (five cases) (two sudden death). i, Right coronary artery from posterior aortic sinus (P) (one case). j, Right coronary artery from above right aortic sinus (two cases). k, Right coronary artery from above right-posterior commissure (two cases). l, Right coronary artery from above right-left commissure (one case). LAD, Left anterior descending coronary artery.

chambers were hypertrophic. The right coronary artery arose from the left aortic sinus and was the dominant artery. It supplied both the atrioventricular and sinoatrial nodes.

Our cases 2 and 3, as far as we are aware, constitute the sixth and seventh reported cases of sudden death in which the right coronary artery arose from the left aortic sinus. It must be acknowledged that in our case 3 the patient's asthmatic condition cannot be excluded as a cause of sudden death, although the evidence favors death caused by another factor.

While sudden death is clearly more common when the left coronary artery arises from the right aortic sinus than when the right coronary artery arises from the left aortic sinus, the latter condition should be recognized as a potential cause of fatal myocardial ischemia. Previous reviews on this subject have noted that sudden death associated with an anomalous coronary artery arising from the aorta usually occurs during or shortly following a period of vigorous exercise.² To our knowledge, most patients whose cases are reported in the English literature are male. Only one previously reported case was a female subject.⁶ Our fatal case 1, that of a 15-year-old girl who died suddenly while playing basketball, is thus the second reported case in a female subject.

There has existed considerable controversy as to the mechanism by which the ectopically arising coronary artery causes sudden death. Proposed theories include distal kinking of the involved artery or relative hypoplasia of the vessel. A popular theory is that the artery of concern is compressed as it passes anteriorly between the aorta and pulmonary trunk. Allegedly, this compressive effect would be intensified during exercise as the aorta and pulmonary artery expand with increased cardiac output. There has been recent angiographic evidence to support this theory.^{7,8}

Although compression may play a role in these cases, we favor the theory proposed by Cheitlin et al;⁵ they suggest that the acute angle at which the ectopic artery leaves the aorta results in a flap-like mechanism at the coronary ostium. Expansion of the aorta during exercise causes this flap to obstruct the already slit-like ostium. They also ascribe importance to the intramural course of the ectopic artery causing it to adhere to the wall of the aorta. Each of the three cases of sudden death in our series demonstrated the flap phenomenon, and in one of the cases, an intramural course of the proximal portion of the anomalous coronary artery was present.

In the study of Virmani et al,¹¹ evidence is presented that peculiar angulation of nonectopically arising coronary arteries may, as in ectopically arising coronary arteries, be a basis for coronary arterial stenosis and of sudden death. The theory of Cheitlin et al⁵ is also strengthened by reports of myocardial infarction in

patients with nonatherosclerotic ectopic coronary arteries which passed either posterior to the aorta or anterior to the pulmonary trunk and thereby could not be compressed between the great vessels.¹²⁻¹⁴

We suggest that any ectopically arising coronary artery that leaves the aorta at an acute angle may be a cause of ischemic heart disease as manifested by angina pectoris, myocardial infarction, or sudden death. Although uncommon, this condition must be considered in the clinical setting of the young patient with symptoms suggestive of myocardial ischemia. Corrective surgery, either coronary arterial bypass grafting or plastic revision of a coronary ostium might be lifesaving.^{7,15}

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