

Coronary Artery Anomalies

A Review of More than 10,000 Patients from
The Clayton Cardiovascular Laboratories

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We reviewed the records of 10,661 patients who had undergone coronary angiography at the Clayton Foundation Cardiovascular Laboratories between 1 June 1974 and 15 March 1986, and identified major coronary artery anomalies in 83 adults. In addition, we included in our review 9 adults and 2 adolescents who had been referred for evaluation of anomalies documented elsewhere. Here we present the clinical and angiographic data for all 94 patients (76 men and 18 women). Most patients were men who presented with chest pain. The most common anomaly, found in 38 patients, was origin of left circumflex coronary artery from right coronary artery or right aortic sinus. In contrast to other studies, which have not shown increased incidence of coronary atherosclerosis in the anomalous circumflex artery, 71% of our patients with this anomaly had significant coronary atherosclerosis in the proximal portion of the anomalous vessel. The posterior course of the anomalous circumflex coronary artery may predispose this vessel to atherosclerosis in patients with coronary disease. The overall incidence of atherosclerotic disease in coronary arteries was 68% (64 of 94 patients) in the present study. (Texas Heart Institute Journal 1988;15:166-173)

Key words: *Coronary vessel anomalies; coronary vessels/abnormalities; heart defects, congenital; cross sectional studies; coronary vessels/radiography; coronary arteriosclerosis; cineangiography; angina pectoris; arteriovenous fistula; coronary disease*

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Major or minor congenital anomalies of the coronary arteries are present in about 2% of adults undergoing cardiac catheterization.¹ Depending upon the origin, course, and termination of the anomalous vessel, certain coronary anomalies may be associated with sudden death, syncope, other congenital heart disease, or anginal syndromes, or they may be incidental findings without adverse prognoses. Accurate recognition and documentation of coronary artery anomalies at the time of coronary angiography are essential to determine the significance of such findings and to avoid therapeutic complications.

The incidence of various coronary anomalies and associated clinical, angiographic and hemodynamic findings have been cited in several clinical series.²⁻⁶ Comprehensive classifications of coronary anomalies have been proposed and some of them accepted.⁷⁻⁹ To compare our experience with previously reported studies, we have reviewed the clinical and angiographic findings for 94 patients with coronary artery anomalies.

Patients and Methods

We surveyed retrospectively the records of 10,661 patients who had undergone coronary angiography at the Clayton Foundation Cardiovascular Laboratories between 1 June 1974 and 15 March 1986, and identified 83 adults with major coronary artery anomalies. In addition, we included 9 adults and 2 adolescents who had been referred for evaluation of coronary anomalies documented elsewhere by means of coronary angiography. These 94 patients (76 men and 18 women) displayed 95 coronary artery anomalies. Clinical characteristics of each patient had been recorded at the time of catheterization, and their subsequent hospital courses charted. Selective coronary angiography had been performed by the Judkins femoral or Sones brachial technique, and coronary cineangiograms filmed with a 35-mm camera at 60 frames/sec, using a 6-inch image intensifier. The images were studied by at least 2 experienced angiographers, who grouped anomalies according to the classification scheme of Vloder, Neufeld, and Edwards:⁸ 1) both coronary arteries arise from the left aortic sinus; 2) both coronary arteries arise from the right aortic sinus; 3) the circumflex artery (Cx) arises from the right sinus of Valsalva or right coronary artery (RCA); 4) the left anterior descending artery (LAD) arises from the right sinus of Valsalva or the RCA; 5) only

TABLE I. Distribution of Anomalies and Associated Diseases and Conditions in 94 Patients Displaying 95* Major Coronary Artery Anomalies

Anomalies	Number of Anomalies	CAD in Anomalous Vessel	CAD in Other Vessels	Valve Disease	Other Abnormalities
Both coronary arteries from RSV	3	-	2	-	-
Both coronary arteries from LSV	30	2	23	3 AS	1
Cx from RCA or RSV	38	27	23	5 AS 2 MVP	2
LAD from RCA	2	-	1	1 MVP	1 TOF
Single CA	7	4	-	-	-
LCA from PT	3	-	-	-	-
LAD from PT	4	-	2	-	1
CA fistulas	8	-	3	-	2 TOF
TOTALS	95	33	54	11	7

*One patient, a 56-year-old woman, had two anomalies: an LAD arising from the PT and a Cx arising from the RCA. She had no associated CAD.

AS = aortic stenosis; CA = coronary artery; CAD = coronary artery disease; Cx = circumflex coronary artery; LAD = left descending coronary artery; LCA = left coronary artery; LSV = left sinus of Valsalva; MVP = mitral valve prolapse; PT = pulmonary trunk; RCA = right coronary artery; RSV = right sinus of Valsalva; TOF = tetralogy of Fallot

1 coronary artery exists; and 6) a coronary artery communicates anomalously with a cardiac chamber or major thoracic vessel.

In this report, we consider only major anomalies. Several minor anomalies were not considered. Excluded from evaluation were those cases wherein: 1) the coronary ostium was above the sinotubular junction of the appropriate sinus of Valsalva (origin of the coronary ostium above the junctional line is considered a minor variation occurring in about 8% of adult hearts⁸); 2) the Cx and LAD originated from separate ostia in the left sinus of Valsalva (an anomaly found in about 1% of autopsy cases⁸); 3) the RCA displayed separate ostia of proximal branches; 4) the RCA originated from an unusual site within the right sinus of Valsalva; and 5) a minor communication occurred between the left coronary artery (LCA) and pulmonary artery.

Results

Among the 94 patients, 95 coronary artery anomalies were identified (Table I). Sixty-four patients (68%) had associated coronary artery disease (defined as more than 50% luminal stenosis of 1 or more major epicardial coronary arteries). Eighteen patients (19%) had cardiac abnormalities other than coronary artery disease.

Origin of Both Coronary Arteries from Right Aortic Sinus

The LCA originated from the right aortic sinus in 3 patients. Two were men, ages 51 and 53 years, and 1 was a 76-year-old woman. None had a history of syncope or prior myocardial infarction. All 3 patients had cardiac catheterization and coronary angiography for evaluation of chest pain. Figure 1A is a diagram showing the origin and course of normal coronary arteries. The initial course of the LCA in these 3 patients was between the aorta and pulmonary artery (Fig. 1B). The course and distribution of the RCA were normal in all 3 patients. Significant coronary artery disease was identified in 2 patients, but no stenosis was present in their anomalous LCAs.

Origin of Both Coronary Arteries from the Left Aortic Sinus

The RCA arose from the left aortic sinus in 30 patients, beginning anteriorly to the left main coronary artery (LM) and coursing anteriorly between the aorta and pulmonary artery (Fig. 1C). Twenty-two of these patients were men and 8 were women; the mean age was 59.9 years. Twenty-six patients were evaluated for chest pain, and 23 were found to have significant coronary atherosclerosis. Two patients (6.7%) had coronary artery disease involving the anomalous portion of the proximal RCA: 1 had a single 99%

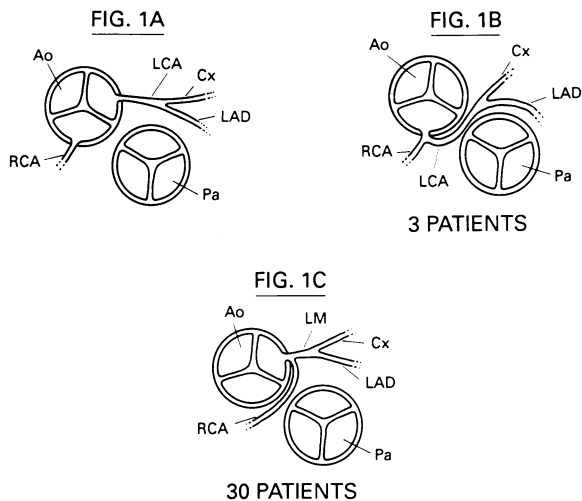


Fig. 1 Diagrams showing **A)** origin and course of normal coronary arteries, **B)** both coronary arteries originating from the right sinus of Valsalva, and **C)** both coronary arteries originating from the left sinus of Valsalva.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; LCA = left coronary artery; LM = left main coronary artery; Pa = pulmonary artery; RCA = right coronary artery

occlusion of this segment, while the other had a 95% occlusion of the proximal anomalous RCA and high grade stenoses of his LM and Cx. Three patients had hemodynamically significant aortic stenosis: 1 of these patients had a bicuspid valve documented at surgery, and another cited a history of syncope. No other patients had a history of syncope.

Anomalous Origin of the Circumflex Coronary Artery

Anomalous origin of the Cx from the right aortic sinus or the 1st portion of the RCA was observed in 38 patients. Thirty-three patients were men and 5 were women (mean age, 55.4 years). In all cases, the initial course of the Cx was posterior to the aorta (Figs. 2A and 2B). Coronary artery disease was found in 27 patients (71%), and 21 patients were found to have significant stenosis of all 3 of their coronary arteries. All 27 patients with coronary artery disease had significant stenosis of the proximal portion of their anomalous Cx vessels. Four patients presenting with typical angina pectoris had stenosis of the anomalous vessel alone. Hemodynamically significant aortic stenosis was present in 5 patients, and an atrial septal defect was documented in another. Of the patients with aortic stenosis, 4 showed no evidence of prior rheumatic fever, and 2 had bicuspid aortic valves documented at surgery.

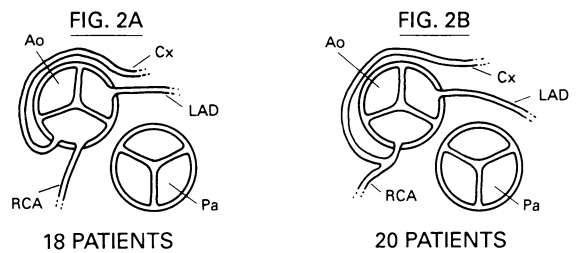


Fig. 2 Diagrams showing **A)** origin and course of anomalous circumflex artery arising from right aortic sinus and **B)** from right coronary artery.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; Pa = pulmonary artery; RCA = right coronary artery

Anomalous Origin of the Left Anterior Descending Artery from the Right Aortic Sinus

The LAD originated from the right aortic sinus in 2 patients. Both were men, ages 46 and 69 years. One had angina pectoris as a result of single-vessel coronary artery disease and the other had tetralogy of Fallot. In both patients, the initial course of the LAD was anterior to the right ventricular outflow tract (Fig. 3). Neither patient had a history of syncope nor of myocardial infarction.

Single Coronary Artery

Single coronary artery was observed in 7 patients (6 men and 1 woman), whose mean age was 58.2 years. In 4 of these patients, the initial portion of the single coronary artery followed the path of a normal LCA but continued posteriorly in the atrioventricular groove to the area of the heart normally supplied by the RCA

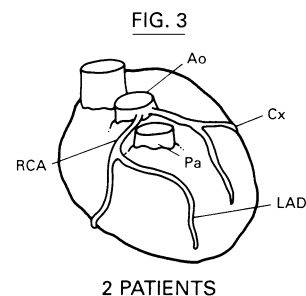


Fig. 3 Diagram of origin and course of anomalous left anterior descending artery arising from the right coronary artery.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; Pa = pulmonary artery; RCA = right coronary artery

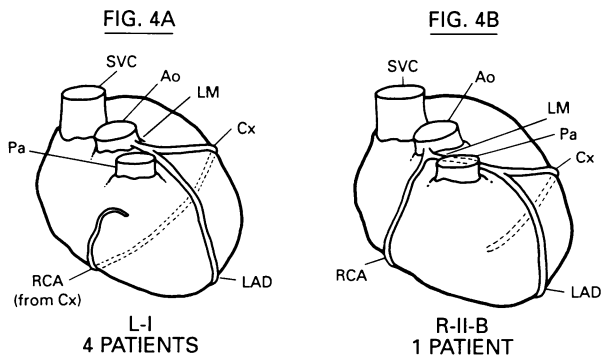


Fig. 4 Diagrams of **A**) L-I type single coronary artery branching posteriorly (Cx) in the atrioventricular groove and **B**) R-II-B type single coronary artery with a single large trunk (LM) coursing between the aorta and pulmonary artery.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; LM = left main coronary artery; Pa = pulmonary artery; RCA = right coronary artery; SVC = superior vena cava

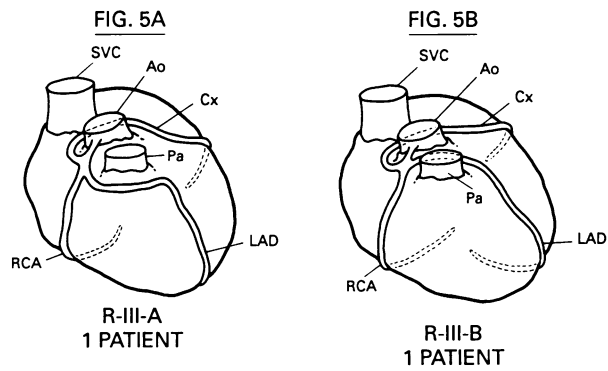


Fig. 5 Diagrams of **A**) R-III-A type single coronary artery, showing the course of the left anterior descending artery anterior to the pulmonary artery, and **B**) R-III-B type single coronary artery, showing the course of the left anterior descending artery between the aorta and pulmonary artery.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; Pa = pulmonary artery; RCA = right coronary artery; SVC = superior vena cava

(Fig. 4A). These 4 patients were classified as having an L-I anomaly, according to the system defined by Lipton and colleagues.¹⁰ Another patient's single coronary artery arose from the right coronary sinus, with a single large trunk traversing between the aorta and pulmonary artery to areas normally supplied by the Cx and LAD (Fig. 4B). This was classified as an R-II-B anomaly.¹⁰

In the remaining 2 patients, the single coronary artery arose from the right coronary sinus and trifurcated, with the Cx passing posteriorly and the LAD passing anteriorly to the pulmonary artery (Fig. 5A) or between the great vessels (Fig. 5B). These single coronary arteries were classified as R-III-A and R-III-B anomalies.¹⁰

No associated cardiac abnormalities were observed, except for coronary artery disease in 4 patients. Two of the 4 patients who displayed an L-I anomaly had significant stenosis of the LAD or of the proximal single coronary artery (prior to the point of bifurcation); both had presented with congestive heart failure and evidence of prior myocardial infarction. Both patients with R-III anomalies had significant stenosis of their posteriorly coursing Cx arteries.

Anomalous Communication or Origin of a Coronary Artery with a Cardiac Chamber or Major Thoracic Vessel

The LCAs of 3 patients originated from their main pulmonary arteries (Fig. 6A). Two were men, ages 16 and 57 years; 1 was a 33-year-old woman. All 3 displayed resting and exertional chest pain. None had electrocardiographic evidence of prior myocardial in-

farction. At coronary angiography, all 3 were observed to have dominant RCAs and well-developed collateral vessels with retrograde flow to their main pulmonary arteries via their LCAs.

In 4 patients, the LAD arose from the pulmonary trunk (Fig. 6B). Two patients, a 56-year-old man and a 71-year-old woman, had significant coronary artery disease in addition to the coronary anomaly. Both presented with chest pain. The 3rd patient, a 56-year-old woman, also had an anomalous origin of the Cx

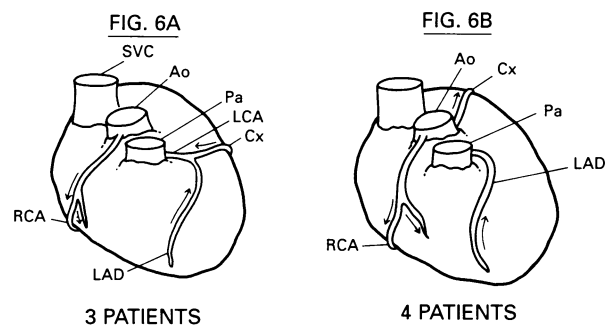


Fig. 6 Diagrams showing **A**) origin of left coronary artery from pulmonary artery, with arrows indicating collateral blood flow from right coronary artery, and **B**) origin of left anterior descending coronary artery from pulmonary artery, with arrows indicating collateral blood flow from right and circumflex coronary arteries.

Ao = aorta; Cx = circumflex artery; LAD = left anterior descending artery; LCA = left coronary artery; Pa = pulmonary artery; RCA = right coronary artery; SVC = superior vena cava

from the right aortic sinus. She had electrocardiographic evidence of a prior anterior-wall myocardial infarction and presented with chest pain, but coronary angiography did not reveal significant coronary artery disease. The 4th patient was a 17-year-old man who had no symptoms or physical findings except a loud systolic murmur.

Eight patients had fistulous connections between a coronary artery and the pulmonary artery or a cardiac chamber. A 29-year-old man who experienced chest pain while jogging had 2 fistulas between his Cx and main pulmonary artery. Hemodynamic studies at cardiac catheterization were normal. Three men had an abnormal connection between a coronary artery and the right atrium: 2, ages 32 and 57 years, had a Cx-to-right atrial fistula. Only 1 of these patients was symptomatic, and in him we found a 2:1 left-to-right shunt at cardiac catheterization. The 3rd man, 60 years old, had an RCA-to-right atrial fistula, but no symptoms. The other 4 patients had abnormal connections between 1 or more coronary arteries and the left ventricle. Two patients with tetralogy of Fallot, a man and woman, both 28 years of age, had multiple small fistulas between the LAD and the left ventricle. The other 2 patients, a 60-year-old man and a 63-year-old woman, had fistulas between both the right and the left coronary arteries and the left ventricle. All patients were observed to have large, dilated coronary arteries when selective coronary angiography was performed.

Discussion

The incidence of coronary artery anomalies in our review is 0.78% (83 of 10,661), which approximates the incidence from other studies of patients referred for coronary angiography.^{1,5} In our study, as in others,^{3,5} coronary artery anomalies appear to be more common in men than in women, although this finding and the 68% incidence of coronary artery disease in our patients may reflect the selective nature of referral for cardiac catheterization and coronary angiography.

Anomalous origin of both coronary arteries from the left sinus of Valsalva was found in 30 patients (0.28%). The incidence of this anomaly in the general population is unknown because the finding is easily overlooked at autopsy. Because patients with the anomaly are more likely to have symptoms of myocardial ischemia, it is discovered more frequently on angiography than is anomalous origin from the right sinus of Valsalva.⁹ Until 1980, this finding was thought to be a minor congenital anomaly of no clinical significance.² Since then, however, researchers have found associations between this anomaly and acute myocardial infarction, angina pectoris, syncope, ventricular tachycardia, ventricular fibrillation, and

sudden death, in the absence of atherosclerotic or other cardiac disease.¹¹⁻¹⁵ Sudden death, however, is rarely the initial manifestation of this anomaly: it is most often preceded by symptoms of ischemia. On the basis of angiographic studies, it has been estimated that approximately one-third of all patients with this anomaly will have symptoms of myocardial ischemia or dysfunction.⁹ Myocardial ischemia in association with this anomaly is thought to be caused by an abnormal slit-like RCA ostium, acute angulation of the RCA, and compression of the RCA between the aorta and pulmonary trunk during exercise.^{9,11-15} Operative therapy in the form of aortocoronary bypass grafting of the anomalous vessel is not indicated unless symptoms are present.⁹ Twenty-six of our 30 patients had chest pain, and 25 were found to have significant coronary artery disease. Coronary artery disease of the anomalous portion of the RCA has not been previously reported but was found in 2 of our patients. Both had severe stenosis of their proximal RCAs; 1 had no other evidence of coronary atherosclerosis. One patient with hemodynamically significant aortic stenosis had a history of syncope.

Origin of both coronary arteries from the right sinus of Valsalva is found in 0.06% to 0.19% of patients undergoing coronary angiography.¹⁵ In our study, this anomaly was identified in only 3 patients, or 0.03%. Four possible courses of the left coronary artery have been reported: anterior to the pulmonary trunk, posterior to the aorta, within the intraventricular septum beneath the right ventricular outflow tract, and between the aorta and pulmonary trunk.¹⁶ With 1 exception, no cases of myocardial ischemia or sudden death have been reported in association with the anterior, posterior, or septal courses of the LCA in the absence of concomitant coronary artery disease.⁹

In contrast, origin of the LCA from the right sinus with a course between the aorta and the pulmonary trunk is associated with sudden death in the absence of other cardiac abnormalities.^{6,17} In Cheitlin's series,⁶ 27% of patients with this anomaly died suddenly, either during or immediately after exercise. Most sudden deaths occurred in young men. Barth and Roberts¹⁷ reported similar findings in their review of 5 patients with this anomaly and summary of previously reported patients. The significance of this anomaly in patients 20 years of age or older is not clear. One case, also reported by Barth and Roberts,¹⁷ appears to be the only instance in the literature of fatal myocardial ischemia past the 2nd decade, due to this anomaly.

None of our patients with origin of both coronary arteries from the right sinus of Valsalva had symptoms originating from their coronary anomaly. Most young patients who die suddenly with this anomaly, however, do have prodromal ischemic symptoms. Ischemia is thought to be caused by compression of the

LCA between the aorta and pulmonary trunk during exercise, a spasm or kinking of the LCA, acute angulation of the LCA, or an anatomic abnormality at the orifice of the ostium.^{6,9,17,18} Noninvasive evaluation of these symptoms is usually unrewarding, with the exception of echocardiography, which can exclude aortic stenosis or hypertrophic cardiomyopathy. Roberts⁹ recommends coronary angiography if the results of a stress electrocardiographic study are abnormal, or if symptoms persist despite a normal stress electrocardiogram. Operative therapy for this anomaly consists of bypass grafting of both the LAD and Cx, or enlargement of the abnormal left coronary ostium.^{9,19}

Anomalous origin of the Cx from the RCA or the right sinus of Valsalva is the most common coronary anomaly reported in angiographic series and necropsy studies.^{9,20} It was found in 0.48% of our patients. This anomaly is thought to be of little clinical significance unless valve surgery or coronary artery bypass surgery is performed without previous detection of the anomaly, or unless severe atherosclerotic narrowing is present in the RCA proximal to the origin of the Cx.²⁰⁻²² In our patients, this anomaly was associated with valvular heart disease, although the incidence was lower than that noted in other studies.^{4,5} According to Page and coworkers,²⁰ there is no increased incidence of coronary atherosclerosis in the anomalous portion of the Cx. However, 71% of our patients with this anomaly had significant stenosis of the proximal Cx, and 11% had severe atherosclerosis in this vessel alone. It appears that in patients with coronary artery disease, there is a predilection for the development of atherosclerosis in the posteriorly coursing anomalous vessel. Kimbiris and associates⁵ observed similar findings in 2 patients with LCAs originating from the right sinus of Valsalva and coursing posteriorly.

Anomalous origin of the LAD from the RCA or the right sinus of Valsalva is rare in the absence of other cardiac abnormalities.²³ It occurred in 0.02% of our patients. Kimbiris and colleagues⁵ reported an incidence of 0.03%. The course of the LAD is either anterior to the right ventricular outflow tract or through the interventricular septum at the level of infundibulum. This anomaly is frequently associated with tetralogy of Fallot, and vessels have been divided inadvertently during corrective procedures.²⁴

Single coronary artery is a rare anomaly, occurring in 0.06% of our patients. The majority of patients younger than 20 years of age present with an associated abnormality—most frequently transposition of the great vessels or coronary artery fistula—while older patients have a low incidence of associated anomalies.⁹ In the absence of significant coronary atherosclerosis, a single coronary artery may be a benign finding unassociated with functional or

anatomic evidence of ischemia.²⁵ In our series, 2 patients with an R-III¹⁰ single coronary artery had significant atherosclerosis of their posteriorly coursing Cx arteries. This suggests that a posteriorly directed coronary artery associated with the origin of both coronary arteries from the right sinus of Valsalva, with an anomalous origin of the Cx, or with an R-III single coronary artery, is subjected to unusual stress, predisposing it to development of atherosclerosis.

Origin of the LCA from the pulmonary trunk is a rare finding in adults who are undergoing coronary angiography for suspected ischemic heart disease.⁹ Seventy-five percent of patients with this anomaly develop symptoms of congestive heart failure or myocardial ischemia in the 1st 4 months of life, and most die within 2 years of the onset of symptoms.²⁶ Survival is dependent upon the development of collateral channels between the normal RCA and the anomalous LCA, since flow is retrograde in the left system. The older surviving patient may have an abnormal electrocardiogram showing anterior ischemic changes or infarction, angina pectoris, syncope, dyspnea, abnormal stress electrocardiography, a continuous murmur, a mitral regurgitant murmur, sudden death, or no symptoms.²⁶⁻²⁸ Surgical therapy for this anomaly is ligation of the LCA at the pulmonary trunk alone, ligation combined with aortic implantation of the LCA, or saphenous vein bypass grafting to the LAD. Moodie and associates²⁷ report that no data suggest superiority of 1 approach over the others, and that survival is related to the degree of preoperative left ventricular dysfunction.²⁷ However, Grace and coworkers²⁹ indicate that aortic implantation is the procedure of choice. Infants undergoing simple ligation have a higher mortality rate than do infants undergoing the other corrective procedures, and saphenous vein grafts frequently obstruct, or fail to grow with the child.²⁸ All 3 patients in our series had implantation of the LCA to the aorta and experienced uneventful postoperative courses.

Anomalous origin of the LAD from the pulmonary artery was identified in 4 patients (0.04%). Here too, survival past infancy appears to be related to the development of adequate collateral channels from both the RCA and Cx to the LAD.⁹ Older patients with this anomaly may be asymptomatic; but sudden death, myocardial infarction, angina pectoris, and mitral regurgitation have been reported.^{30,31} Even asymptomatic patients with this anomaly should undergo ligation of the LAD at the pulmonary trunk, or ligation combined with a saphenous-vein bypass graft between the aorta and LAD. Reimplantation of the LAD to the aorta is technically inadvisable.³¹

Congenital coronary artery fistulas are uncommon findings in adults undergoing coronary angiography. The majority of fistulas originating from coronary arteries terminate in the right side of the heart, and are

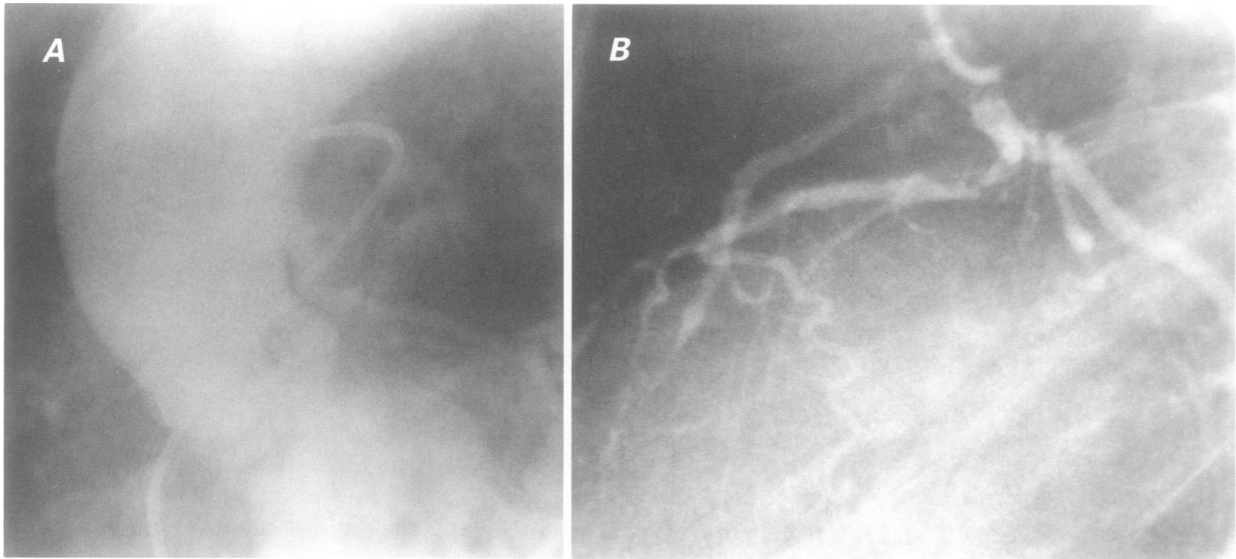


Fig. 7 **A)** An aortogram shows the origin of a single coronary artery from the aorta, above the left sinus of Valsalva; the right and left anterior descending arteries and the circumflex coronary artery can also be seen. **B)** In the same patient, an aortogram in left anterior oblique projection—after selective injection into the single anomalous coronary artery—shows all branches in normal location.

true left-to-right shunts. Unilateral coronary artery fistulas are caused by an embryonic defect in which primitive intratrabecular spaces persist, allowing communication between the developing coronary artery and the cardiac chamber.³² Approximately one-half of the patients with this anomaly develop complications, which include congestive heart failure, sub-acute bacterial endocarditis, myocardial ischemia, and rupture of an aneurysmal fistula.²⁶ Surgical ligation of the fistula in the symptomatic patient is indicated, but surgical therapy for the asymptomatic patient is controversial. Complications

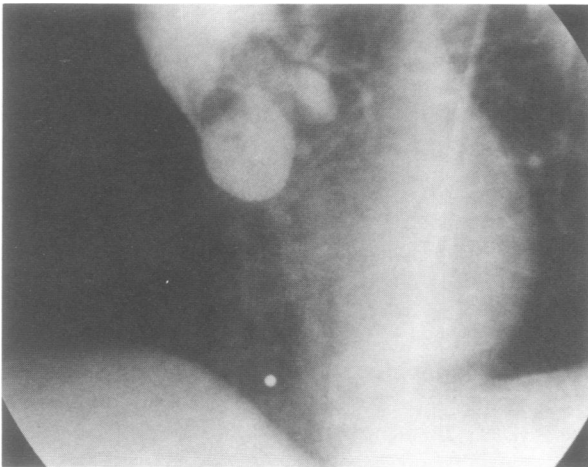


Fig. 8 An aortogram shows an anomalous left coronary artery arising above the left sinus of Valsalva.

in the asymptomatic patient increase with advancing age.³³ Bilateral coronary artery fistulas, extremely rare, are unique in their tendency to terminate in the pulmonary artery. Unlike unilateral coronary fistulas, bilateral fistulas to the pulmonary artery develop by supernumerary implantation of developing coronary arteries into the pulmonary arterial portion of the embryonic truncus arteriosus.³³ More than one-half of all patients with this anomaly become symptomatic, and surgical ligation of fistulas is indicated in patients with left ventricular dysfunction.³⁴

In summary, 0.78% of patients reviewed in our laboratories during a 12-year period were identified as having a congenital coronary anomaly. The majority had been referred for evaluation of chest pain. Anomalous origin of the Cx from the right sinus of Valsalva or RCA was the most common anomaly found. Previous studies have emphasized the benign character of this anomaly.^{4,5,20} Our review suggests that the posterior course of the anomalous Cx appears to predispose this vessel to coronary atherosclerosis. Recognition of coronary anomalies at angiography is mandatory in order to prescribe appropriate medical or surgical therapy.

Addendum

Since the completion of this study, we have encountered in 2 adult male patients a new anomaly not described in this report. In 1 patient, none of the coronary arteries originated from the sinuses of Valsalva. A single coronary artery (Fig. 7A) arose from the ascending aorta approximately 3 cm above the

left sinus of Valsalva. The 1st branch was the RCA, which coursed between the great vessels. The remaining vessel appeared to divide like the LM into the LAD and the Cx, and the branches were in the normal location (Fig. 7B). Coronary artery disease was present in each major branch. In the 2nd patient, the LM originated from the ascending aorta approximately 3 cm above the left sinus of Valsalva (Fig. 8); the branches were normal and the RCA was normal. This patient had associated bicuspid aortic valve with stenosis and regurgitation, but no coronary artery disease.

References

- Baltaxe HA, Wixson D. The incidence of congenital anomalies of the coronary arteries in the adult population. *Radiology* 1977;122:47-52.
- Liberthson RR, Dinsmore RE, Fallon JT. Aberrant coronary artery origin from the aorta. *Circulation* 1979;59:748-54.
- Liberthson RR, Dinsmore RE, Bharati S, et al. Aberrant coronary artery origin from the aorta: diagnosis and clinical significance. *Circulation* 1974;50:774-9.
- Chaitman BR, Lesperance J, Saltiel J, Bourassa MG. Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of the coronary arteries. *Circulation* 1976;53:122-31.
- Kimbiris D, Iskandrian AS, Segal BL, Bemis CE. Anomalous aortic origin of coronary arteries. *Circulation* 1978;58:606-15.
- Cheitlin MD, De Castro CM, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva. *Circulation* 1974;50:780-7.
- Trivellato M, Angelini P, Leachman RD. Variations in coronary artery anatomy: normal versus abnormal. *Cardiovasc Dis Bull Tex Heart Inst* 1980;7:357-70.
- Vlodaver Z, Neufeld HN, Edwards JE. Coronary arterial variations in the normal heart and in congenital heart disease. New York: Academic Press, 1975. 171 p.
- Roberts WC. Major anomalies of coronary arterial origin seen in adulthood. *Am Heart J* 1986;111:941-63.
- Lipton MJ, Barry WH, Obrez I, Silverman JF, Wexler L. Isolated single coronary artery: diagnosis, angiographic classification, and clinical significance. *Radiology* 1979; 130:39-47.
- Benge W, Martins JB, Funk DC. Morbidity associated with anomalous origin of the right coronary artery from the left sinus of Valsalva. *Am Heart J* 1980;99:96-100.
- Roberts WC, Siegel RJ, Zipes DP. Origin of the right coronary artery from the left sinus of Valsalva and its functional consequences: analysis of 10 necropsy patients. *Am J Cardiol* 1982;49:863-8.
- Brandt B III, Martins JB, Marcus ML. Anomalous origin of the right coronary artery from the left sinus of Valsalva. *N Engl J Med* 1983;309:596-8.
- Isner JM, Shen EM, Martin ET, Fortin RV. Sudden unexpected death as a result of anomalous origin of the right coronary artery from the left sinus of Valsalva. *Am J Med* 1984;76:155-8.
- Bett JHN, O'Brien MF, Murray PJS. Surgery for anomalous origin of the right coronary artery. *Br Heart J* 1985;53: 459-61.
- Ishikawa T, Brandt PWT. Anomalous origin of the left main coronary artery from the right anterior aortic sinus: angiographic definition of anomalous course. *Am J Cardiol* 1985; 55:770-6.
- Barth CW III, Roberts WC. Left main coronary artery originating from the right sinus of Valsalva and coursing between the aorta and pulmonary trunk. *J Am Coll Cardiol* 1986;7:366-73.
- Kimbiris D. Anomalous origin of the left main coronary artery from the right sinus of Valsalva. *Am J Cardiol* 1985;55:765-9.
- Davia JE, Green DC, Cheitlin MD, De Castro CM, Brott WH. Anomalous left coronary artery origin from the right coronary sinus. *Am Heart J* 1984;108:165-6.
- Page HL, Engel HJ, Campbell, WB, Thomas CS Jr. Anomalous origin of the left circumflex coronary artery: recognition, angiographic demonstration and clinical significance. *Circulation* 1974;50:768-73.
- Roberts WC, Waller BF, Roberts CS. Fatal atherosclerotic narrowing of the right main coronary artery: origin of the left anterior descending or left circumflex coronary artery from the right (the true "left-main equivalent"). *Am Heart J* 1982;104:638-41.
- Fifer MA, Neiterman AJ, Akins CW. Right main coronary artery disease: antemortem diagnosis and treatment. *Am Heart J* 1986;111:787-8.
- Lardani H, Sheldon WC. Ectopic origin of the left anterior descending coronary artery from the right coronary sinus: report of a case simulating anterior descending obstruction. *Chest* 1976;69:548-9.
- Heuser RR, Achuff SC, Brinker JA. Inadvertent division of an anomalous left anterior descending coronary artery during complete repair of tetralogy of Fallot: 22-year follow-up. *Am Heart J* 1982;103:430-2.
- Barbour DJ, Roberts WC. Origin of the right from the left main coronary artery (single coronary ostium in aorta). *Am J Cardiol* 1985;55:609.
- Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries: angiographic aspects. *Circulation* 1978;58:25-34.
- Moodie DS, Fyfe D, Gill CC, et al. Anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome) in adult patients: long-term follow-up after surgery. *Am Heart J* 1983;106:381-8.
- Vesturlund T, Thomsen PEB, Hansen OK. Anomalous origin of the left coronary artery from the pulmonary artery in an adult. *Br Heart J* 1985;54:110-2.
- Grace RR, Angelini P, Cooley DA. Aortic implantation of anomalous left coronary artery arising from pulmonary artery. *Am J Cardiol* 1977;39:608-13.
- Tamer DF, Mallon SM, Garcia OL, Wolff GS. Anomalous origin of the left anterior descending coronary artery from the pulmonary artery. *Am Heart J* 1984;108:341-5.
- Roberts WC, Robinowitz M. Anomalous origin of the left anterior descending coronary artery from the pulmonary trunk with origin of the right and left circumflex coronary arteries from the aorta. *Am J Cardiol* 1984;54:1381-3.
- Pezzella AT, Falaschi G, Ott DA, Cooley DA. Congenital coronary artery-left heart fistulas: report of three cases. *Cardiovasc Dis Bull Tex Heart Inst* 1981;8:355-63.
- Baim DS, Kline H, Silverman JF. Bilateral coronary artery-pulmonary artery fistulas: report of five cases and review of the literature. *Circulation* 1982;65:810-5.
- Babb JD, Field JM. Double coronary arteriovenous fistula. *Chest* 1977;72:656-8.