

Catheterization Curriculum

Coronary Artery Anomalies in 126,595 Patients Undergoing Coronary Arteriography

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Coronary artery anomalies were found in 1,686 patients (1.3% incidence) undergoing coronary arteriography at the Cleveland Clinic Foundation from 1960 to 1988. Of the 1,686 patients, 1,461 (87%) had anomalies of origin and distribution, and 225 (13%) had coronary artery fistulae. Most coronary anomalies did not result in signs, symptoms, or complications, and usually were discovered as incidental findings at the time of catheterization. Eighty-one percent were "benign" anomalies: 1) separate origin of the left anterior descending and circumflex from the left sinus of Valsalva; 2) ectopic origin of the circumflex from the right sinus of Valsalva; 3) ectopic coronary origin from the posterior sinus of Valsalva; 4) anomalous coronary origin from the ascending aorta; 5) absent circumflex; 6) intercoronary communications; and 7) small coronary artery fistulae. Other anomalies may be associated with potentially serious sequelae such as angina pectoris, myocardial infarction, syncope, cardiac arrhythmias, congestive heart failure, or sudden death. Potentially serious anomalies include: 1) ectopic coronary origin from the pulmonary artery; 2) ectopic coronary origin from the opposite aortic sinus; 3) single coronary artery; and 4) large coronary fistulae. Coronary artery anomalies require accurate recognition, and at times, surgical correction.

Key words: anomalous origin of coronary artery, coronary artery from pulmonary trunk, Bland-White-Garland syndrome, single coronary artery, coronary artery fistulae

INTRODUCTION

Normal coronary artery anatomy encompasses a wide spectrum. Minor variations, such as number of septal perforator or diagonal branches and size and distribution of individual vessels, ensure that no two anatomic patterns are exactly alike. Coronary artery anomalies represent marked deviations from the normal pattern. Anomalies are present at birth, but relatively few are symptomatic during childhood. Most anomalies are discovered as incidental findings during coronary arteriography or at autopsy. However, some anomalies present with symptoms or potentially serious sequelae that require surgical treatment. The clinician should suspect the presence of a coronary artery anomaly in a young person who experiences exertional syncope, myocardial infarction, exercise-induced arrhythmias, or cardiac arrest (1–4). The angiographer should be able to identify anomalies in order to perform accurate evaluations and avoid subsequent errors in management. The cardiac surgeon must be aware of the abnormal anatomy in order to avoid accidental ligation or transection at the time of surgery (5–7). This paper defines the anatomical patterns, frequency of occurrence, and clinical significance of coro-

nary artery anomalies in patients studied angiographically.

MATERIALS AND METHODS

The data base consisted of all patients undergoing coronary arteriography at the Cleveland Clinic Foundation from 1960 to 1988. Patients with isolated coronary artery anomalies were identified and served as the study group. Clinical histories, physical examinations, noninvasive laboratory studies, catheterization data, surgical reports, and follow-up surveys on subgroups of patients were obtained. Patients with separate origin of the conus branch or right ventricular branch from the right sinus of Valsalva were excluded. Patients with coronary anoma-

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Received February 10, 1990; accepted May 11, 1990.

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TABLE I. Isolated Congenital Coronary Artery Anomalies

	No.	Incidence (%)	Anomalies (%)
Total coronary arteriograms	126,595		
Total coronary anomalies	1,686	1.33	
Anomalies of origin and distribution	1,461	1.15	87
Coronary artery fistulae	225	0.18	13

lies occurring as part of complex congenital heart disease were not included in this study.

RESULTS

Patient Population

The study included 126,595 patients who underwent coronary arteriography during the 28 year study interval. This was primarily an adult coronary arteriographic population studied to determine whether arteriosclerotic coronary disease was present or absent. The mean age of patients was 53 years with a range of 9 months to 82 years. Seventy-four percent were males and 26% females; 59% had arteriosclerotic heart disease, 9% had rheumatic heart disease, 7% had congenital heart disease, and 6% had miscellaneous heart disease.

Incidence

The study identified 1,686 consecutive patients with isolated congenital coronary artery anomalies (1.3% incidence). Of these, 1,461 patients (87%) had anomalies of origin and distribution, and 225 patients (13%) had coronary artery fistulae (Table I). This series is compared with other previously published reports in Table II.

Classification

Anatomically, patients were classified into two major groups: those with anomalies of origin and distribution, and those with coronary artery fistulae. Clinically, anomalies may be arbitrarily divided into "benign" and "potentially serious" (Table III).

DISCUSSION

This series is a compilation of coronary anomalies in an adult population studied over a 28 year interval. Patient selection may tend to favor benign asymptomatic anomalies since the more malignant anomalies would have been detected earlier in life or resulted in sudden death. All anomalies are present at birth and occur equally in both sexes. Coronary artery disease does not seem to occur with increased incidence in anomalous vessels nor does the presence of coronary anomalies protect the patient against coronary artery disease. The clas-

TABLE II. Incidence of Coronary Artery Anomalies, Angiographic Series

Author (ref.)	Total no. patients	Anomalies	Incidence (%)
Liberthson (8)	Not stated	21	0.6
Engel (9)	4,250	51	1.2
Chaitman (10)	3,750	31	0.83
Baltaxe (11)	1,000	9	0.9
Kimbiris (12)	7,000	45	0.64
Donaldson (13)	9,153	82	0.9
Hobbs (14) ^a	38,703	601	1.55
Wilkins (15)	10,661	83	0.78
Yamanaka	126,595	1,686	1.3

^aThis is an earlier subgroup of this series of patients.

sification of benign versus potentially serious is arbitrary and based on current available information.

Benign Coronary Anomalies

Separate origin of left anterior descending artery (LAD) and left circumflex (CX) from the left sinus of Valsalva (absent left main trunk). In this anomaly, the LAD and CX arise from separate, but adjacent ostia in the left sinus of Valsalva (Fig. 1). The vessels are otherwise normal in their distribution patterns. This anomaly is found with increased incidence in aortic valve disease and dominance of the left coronary artery. (14) Angiographically, it may be difficult at times to determine whether the left main trunk (LMT) is very short or absent. Contrast injection into the left sinus of Valsalva in the LAO caudal projection provides the best view for making the distinction. If the cardiac catheter is positioned anteriorly, the LAD alone will be visualized and left lateral wall will appear nonperfused. Likewise, when the catheter is positioned posteriorly, the CX alone will be visualized. Occasionally, the anomaly is not recognized at the time of catheterization, and the LAD or CX may be misinterpreted as totally obstructed or congenitally absent. An error in angiographic diagnosis might in turn lead to an error in clinical management. This anomaly causes no hemodynamic impairment and should be considered "benign."

Absent left circumflex. In this anomaly, a large "superdominant" right coronary artery (RCA) crosses the crux of the heart where it ascends in the AV groove and perfuses the posterolateral and lateral wall of the heart (Fig. 2). The LAD arises from the left sinus of Valsalva and has normal distribution. This anomaly is benign in the absence of coronary occlusive disease.

Origin of left circumflex from right coronary or right sinus of Valsalva. In this very common anomaly, the CX arises from the right sinus of Valsalva or proximal RCA, courses posterior to the aorta, and provides branches to the left lateral wall of the heart (Fig. 3). The

TABLE III. Isolated Coronary Artery Anomalies*

	No.	Incidence (%)	Anomalies (%)
Benign			
Separate origin of LAD and CX in LSV	513	0.41	30.4
CX from RSV or RCA	467	0.37	27.7
Coronary artery from PSV			
LMT from PSV	1	0.0008	0.06
RCA from PSV	4	0.003	0.24
Anomalous origin from ascending aorta			
LMT from aorta	16	0.013	0.95
RCA from aorta	188	0.15	11.2
Absent CX ("super-dominant RCA")	4	0.003	0.24
Intercoronary communication	3	0.002	0.18
Small coronary artery fistulae	163	0.12	9.7
Total	1,359	1.07	80.6
Potentially serious			
Coronary artery from pulmonary artery			
LMT from pulmonary artery	10	0.008	0.59
LAD from pulmonary artery	1	0.0008	0.06
RCA from pulmonary artery	2	0.002	0.12
Coronary origin from opposite aortic sinus			
LMT from RSV	22	0.017	1.3
LAD from RSV	38	0.03	2.3
RCA from LSV	136	0.107	8.1
Single coronary artery ^a			
R-I	1	0.0008	0.06
R-II	19	0.015	1.1
R-III	5	0.004	0.30
L-I	20	0.016	1.2
L-II	11	0.009	0.65
Multiple or large sized fistulae	62	0.05	3.7
Total	327	0.26	19.4

*LAD, left anterior descending; CX, circumflex; RCA, right coronary artery; LMT, left main trunk; LSV, left sinus of Valsalva; RSV, right sinus of Valsalva; PSV, posterior sinus of Valsalva.

^aModified Lipton classification; see text.

LAD and RCA are normal. The anomaly should be suspected when contrast injection into the left coronary artery reveals an unusually long, nonbranching proximal segment and a nonperfused left lateral wall (16). Contrast injection into the RCA may also fail to opacify the anomalous CX or demonstrate collateral vessels to the left lateral wall. If the catheter is repositioned near the ostium or in a posterior location in the right sinus of Valsalva, the anomalous CX will be visualized. Inability to recognize this anomaly may prolong the catheterization or necessitate repeating the study. If the angiographer assumes the vessel is occluded or congenitally absent, clinical mismanagement may result. The cardiac surgeon should be informed of the anomalous left circumflex in order to avoid accidental compression of the vessel during valve replacement (17). Since none of the patients in this series had angina pectoris, myocardial infarction, etc., in the absence of coronary atherosclerosis, the anomaly may be considered "benign".

Ectopic origin of right coronary artery or left main trunk from posterior sinus of Valsalva. These anomalies are extremely rare. Except for the ectopic origin, the anatomical course is normal. In this series, no clinical manifestations or perfusion abnormalities were associated with these anomalies. In the few cases reported in the literature, the anomaly itself was not associated with symptoms or complications (18). Until evidence is presented to the contrary, this anomaly should be considered "benign."

Ectopic coronary origin from the ascending aorta. This anomaly is suspected when the angiographer is unable to locate a coronary artery within the sinuses of Valsalva. A search within the proximal 2 cm of the ascending aorta usually reveals the anomalous orifice. Except for the high estium, distribution is normal (Fig. 4). This anomaly is benign, but the cardiac surgeon should be cautioned to avoid accidentally crossclamping or transecting the vessel during surgery (19).

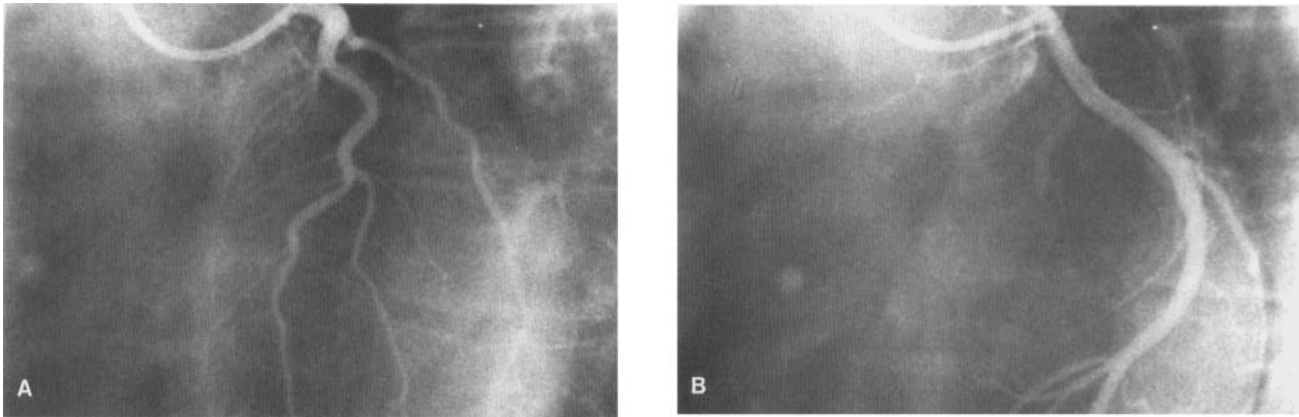


Fig. 1. Separate origin of left anterior descending and circumflex from the left sinus of Valsalva (absent left main trunk). **A:** Anterior angulation of the catheter results in selective injection of the LAD, LAO projection. **B:** Posterior catheter angulation results in selective injection of the CX, LAO projection.

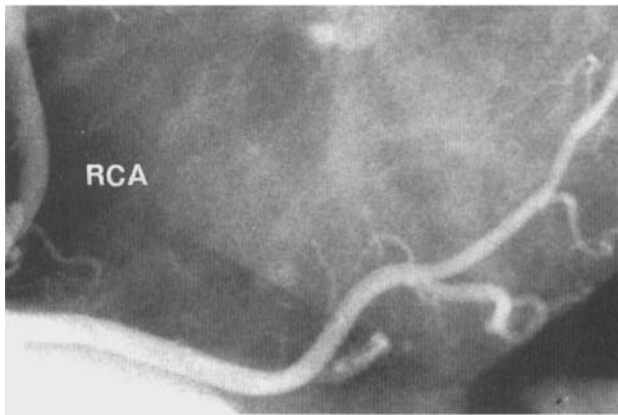


Fig. 2. Absent left circumflex. Selective injection of the right coronary artery (RCA) in the LAO projection demonstrates continuation of the AV branch to the left lateral wall of the heart.

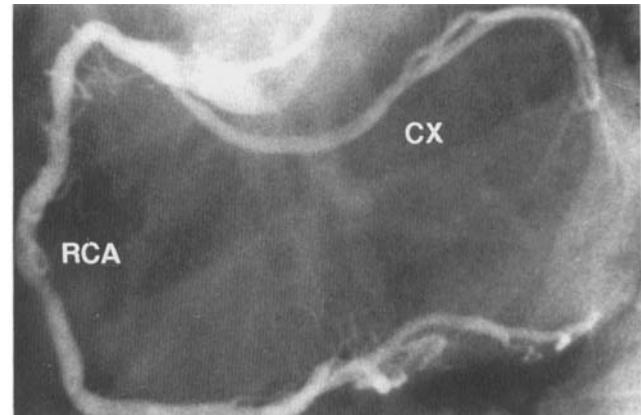


Fig. 3. Ectopic origin of the circumflex from the right coronary artery (RCA), LAO projection. The anomalous circumflex (CX) courses to the left with a caudal-posterior loop. There are mild irregularities of the right coronary artery.

Intercoronary communication. In this rare anomaly, the AV branch of the right and the left circumflex are contiguous, resulting in an open-ended circulation with bi-directional blood flow (Fig. 5). Contrast injection of the RCA fills the CX in a retrograde fashion, and injection of the left coronary artery opacifies the RCA via the AV circumflex branch. A similar communication may also exist between the distal LAD and the distal posterior descending branch. These communications are found in the absence of coronary artery disease, although myocardial perfusion scans occasionally are abnormal at the connecting site. This anomaly may be misinterpreted as a functioning collateral vessel indicative of an unrecognized severe proximal coronary artery obstruction. This anomaly is benign, and may serve as a collateral source if coronary artery obstructions develop (20).

Small coronary artery fistulae. Small coronary artery fistulae are relatively common coronary anomalies. Most are single, i.e., arise from a single branch of coronary artery and drain into a single recipient chamber (Fig. 6). These fistulae generally do not result in signs, symptoms, or complications (21). Continuous murmurs and intracardiac shunts are undetectable in the majority of cases. In this series, more than half of small fistulae originated from the LAD and drained into the pulmonary artery. A subgroup of 101 patients with small fistulae followed clinically up to 11 years did not develop new manifestations or complications attributable to this anomaly (21). Small numbers of patients have undergone serial catheterization studies demonstrating no increase in fistula or shunt size with time. The presence of small

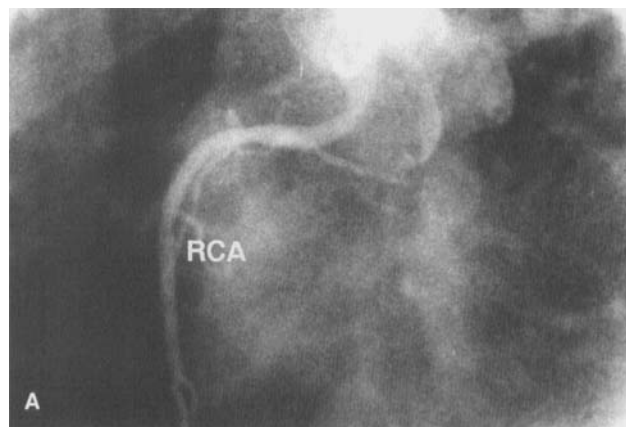
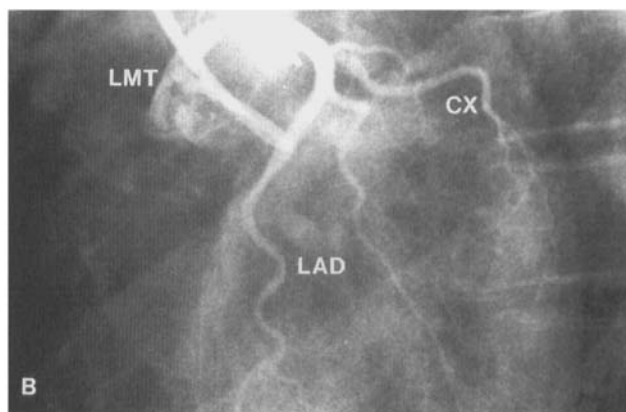


Fig. 4. Ectopic coronary origins from the ascending aorta. A: Right from aorta, LAO projection. Note the vertical orientation of proximal segment. RCA, right coronary artery. B: Left coronary artery from aorta, LAO projection. Note the unusual posi-



tion of the catheter in right anterior aspect of the ascending aorta. CX, circumflex; LAD, left anterior descending artery; LMT, left main trunk.

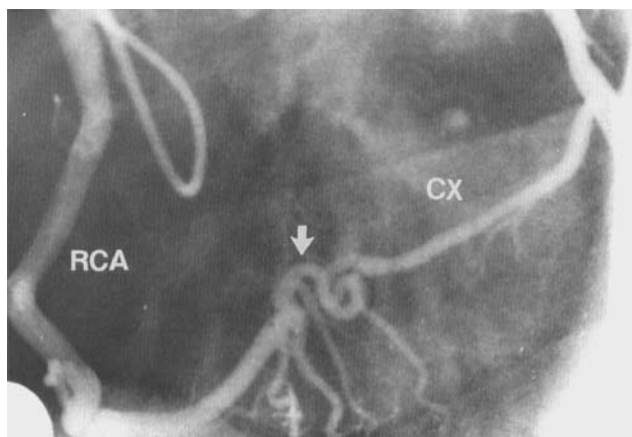


Fig. 5. Intercoronary communication. Selective injection of right coronary (RCA) in LAO projection. The right coronary and circumflex (CX) communicate via an AV branch in the lower center of the frame.

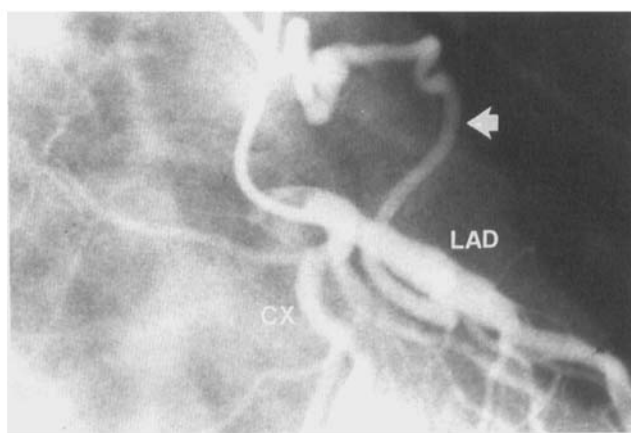


Fig. 6. Small coronary artery fistula. Selective injection of the left coronary in the RAO projection demonstrates a small fistula (arrow), which arises from the LAD and drains into the main pulmonary artery. CX, circumflex.

fistulae in elderly patients attests to their benign nature. Patients with small asymptomatic fistulae should be managed medically.

Potentially Serious Coronary Anomalies

Ectopic coronary origin from the pulmonary artery. Origin of the left coronary artery from the pulmonary artery commonly is referred to as the Bland-White-Garland syndrome (Fig. 7). Blood flow originates in a dilated RCA and passes via collaterals to the left coronary artery where it flows in a retrograde fashion into the pulmonary artery (22). Ninety percent of patients die in infancy and very few survive to reach adulthood (23). Adequate intercoronary collaterals are prerequisite for survival (24). Manifestations include: continuous mur-

mur, angina pectoris, myocardial infarction, mitral regurgitation, congestive heart failure, cardiac arrhythmias and sudden death (25–28). The high risk of sudden death, even in asymptomatic patients, warrants surgical correction of the anomaly when diagnosed (29,30). Many techniques for surgical correction have been utilized: ligation of the left main coronary artery (31,32), left main coronary ligation combined with bypass grafting (31,33), construction of a baffle from the left coronary artery to the aorta (34), left coronary artery to subclavian artery anastomosis (35), re-implantation of the coronary artery into the aorta (31,36), or tunneling from the aorta to left coronary artery using a tissue flap from the pulmonary artery (37,38). The type of surgical procedure may be dictated by the age of the patient and other

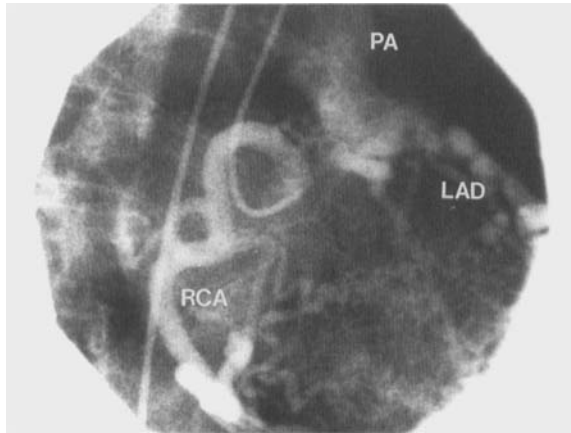


Fig. 7. Ectopic origin of the left main trunk from the pulmonary artery (PA) (Bland-White-Garland Syndrome). Contrast injection in the right coronary artery (RCA) (RAO projection) demonstrates collateral vessels to left coronary (LAD). The left coronary is opacified in a retrograde fashion and connects with the pulmonary artery at the top of the frame.

anatomical factors. Long-term follow-up data is sparse, and reports of vein graft failure with sudden death make the procedure of choice an uncertainty (26).

Origin of the RCA from the pulmonary artery is extremely rare. Blood flows from the left coronary artery via collaterals into the RCA and then retrograde into the pulmonary artery. Although the many patients are asymptomatic, one patient in this series had angina pectoris, and several patients have been reported with cardiac syncope, congestive heart failure, or sudden death (39–41). Ectopic origin of the LAD from the pulmonary artery is the rarest subtype. Contrast injection into either the RCA or left main trunk fills the anomalous LAD via extensive collaterals. In this series, one patient with this anomaly had angina pectoris and evidence of a remote anterior myocardial infarction.

Although color-flow doppler echocardiography may be useful in detecting abnormal pulmonary arterial flow (42), selective coronary arteriography is required for diagnosis. Patients with ectopic origin of a coronary artery from the pulmonary artery should be treated surgically.

Ectopic origin of the left coronary artery from the right sinus of Valsalva. In this anomaly, the entire left coronary artery arises from the right sinus of Valsalva. The RCA may arise separately, or share a common ostium with the anomalous left coronary and is considered a form of single coronary artery. Five anatomical subtypes exist and are classified according to the relationship of the anomalous coronary artery with the aorta and pulmonary artery, i.e., “anterior,” “between,” “septal,” “posterior,” and “combined.” In this series, the “septal” subtype was the most common, whereas the “between” type was rare.

The most potentially serious variant occurs when the left main passes between the aorta and the pulmonary artery prior to its bifurcation (Fig. 8A,B). Angiographically, in the RAO projection, the left main forms a cranial-posterior loop in its initial course between the aortic root and the right ventricular infundibulum at the pulmonic valve level (43). The left main divides in the proximal septum, and the LAD and CX appear to have normal length. A ventriculogram or aortogram performed in the RAO projection will place the LMT on end, giving the appearance of a “dot” on the anterior aspect of the aorta (44). Clinical manifestations include: angina pectoris, syncope, myocardial infarction, ventricular tachycardia, cardiac arrest, and sudden death in the absence of coronary atherosclerosis (8,12,45–51). Symptoms occur mainly in young individuals during physical exertion. (1,3,4) It is postulated that exercise results in the expansion of the aorta, which occludes the acutely angulated slit-like orifice of the left main (45). Older patients frequently are asymptomatic and have normal exercise perfusion scans. Symptomatic patients have undergone coronary artery bypass grafting with good results (4).

The “septal” type is the most common and is called the “intramyocardial” or “tunneled” anomalous variation (Fig. 8C,D). The left main crosses the superior aspect of the crista supraventricularis, passes a variable distance through the septum, and then assumes an epicardial position. This has the appearance of a “hammock” with a caudal-anterior loop seen angiographically (43). The left main becomes epicardial at the mid-septum. The CX courses in a postero-cranial direction, and the LAD appears to be relatively short. The LMT and CX form an ellipse similar to an “eye” with the LMT inferiorly and CX superiorly. Occasionally, a septal perforator will arise from the LMT (44). Most patients are asymptomatic in the absence of coronary disease in other vessels (52). Although this variant frequently is confused with the left main passing between the aorta and pulmonary artery, it is probably a benign anomaly.

In the third anatomical subtype, the left main passes anterior to the pulmonary artery (Fig. 8E,F). The anomalous vessel follows a pattern similar to the conus branch with a cranial-anterior loop (43). The LMT then divides at the junction of the proximal and middle thirds of the septum. In the RAO projection, the LAD appears relatively short and the CX courses posteriorly and inferiorly forming the lower portion of the “eye” with the LMT forming the superior aspect (44). This anomaly usually is benign, although instances of angina pectoris and myocardial infarction in the absence of coronary atherosclerosis in this subtype have been reported (49).

In the fourth pattern, the left main trunk passes posterior to the aorta having a caudal-posterior loop (Fig.

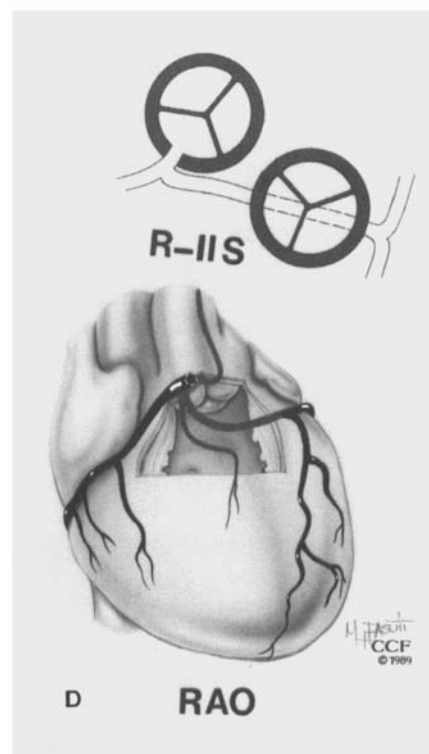
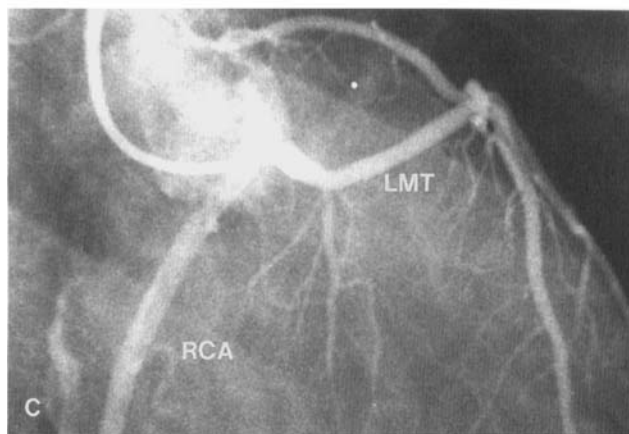
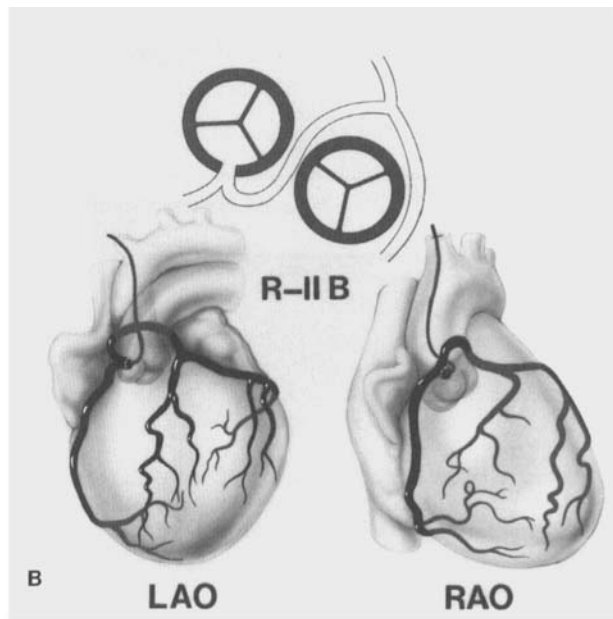
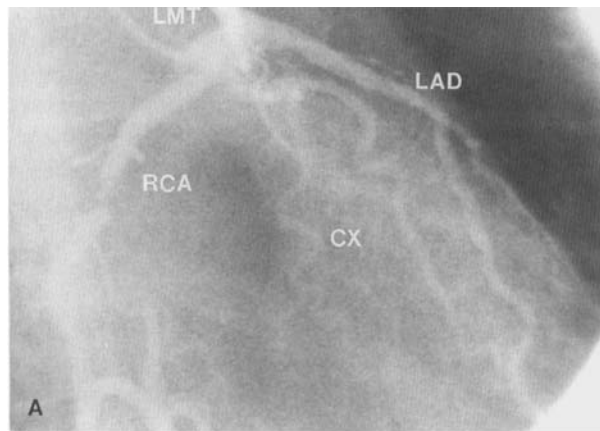


Fig. 8. A,B: Ectopic origin of left coronary artery from the right, "between" type, RAO projection. The left main arises ectopically from the right coronary (RCA) and passes between the aorta and pulmonary artery before dividing into LAD and CX. The LAD and CX appear to have normal length. There is a 60% stenosis in the mid-right coronary artery. **C,D:** Ectopic origin of

left coronary artery from right, "septal" type, RAO projection. The left main trunk (LMT) passes intramurally through the septum, giving off a major septal branch before reaching an epicardial position. This anomaly has a "hammock" appearance with a caudal-anterior loop. RCA, right coronary artery. Figure continued on next page.

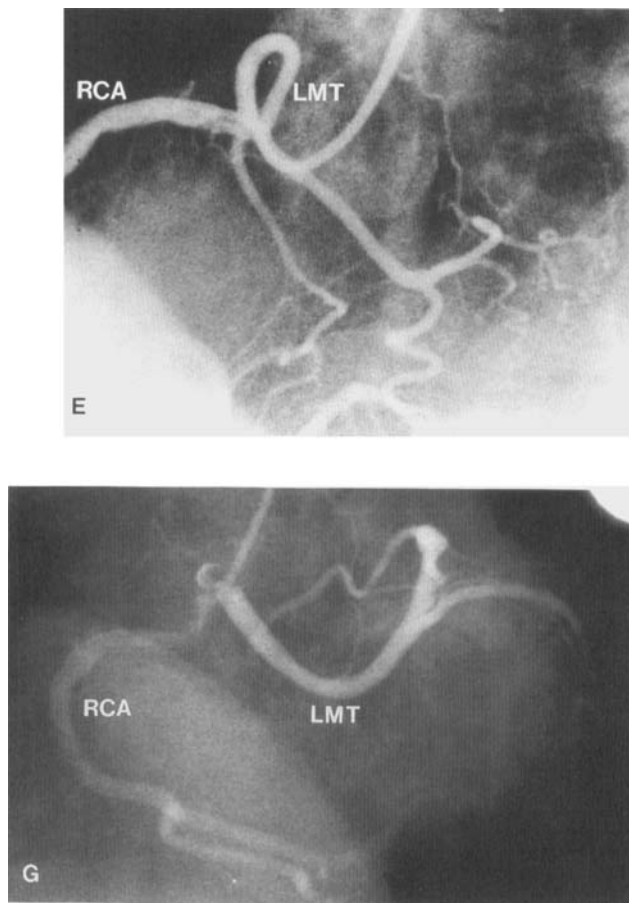
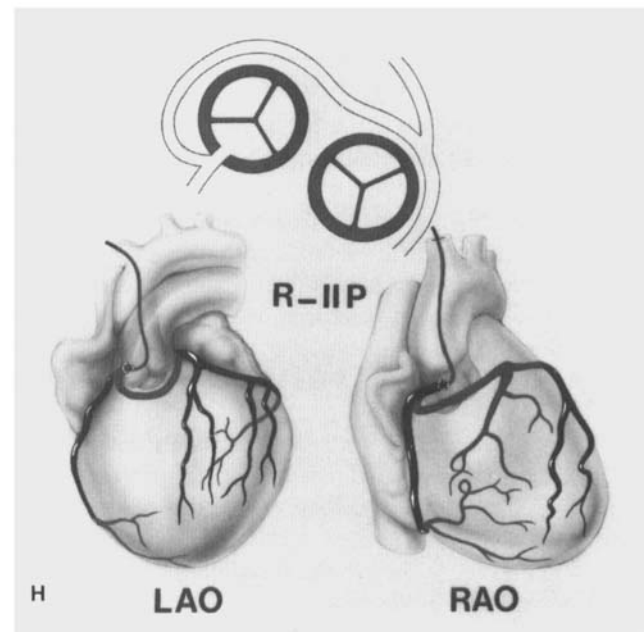
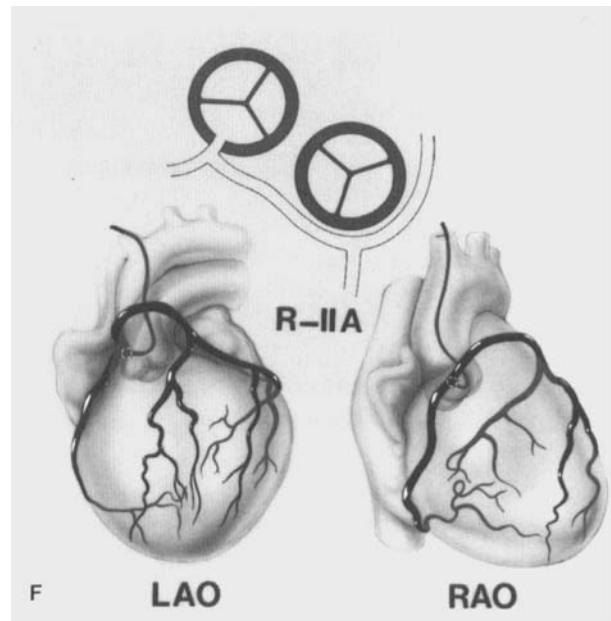


Fig. 8E-H. E,F: Ectopic origin of left coronary artery from right, "anterior" type, LAO projection. The left main trunk (LMT) passes epicardially across the right ventricular outflow tract before dividing into the LAD and CX. This anomaly is similar to conus branch in its initial portion, with a cranial-anterior loop. RCA, right coronary artery. G,H: Ectopic origin of left main trunk (LMT) from right, "posterior" type, LAO projection. The leftmain passes posterior to the aorta with a caudal-posterior loop before dividing into the LAD and CX. RCA, right coronary artery. Figure continued on next page.

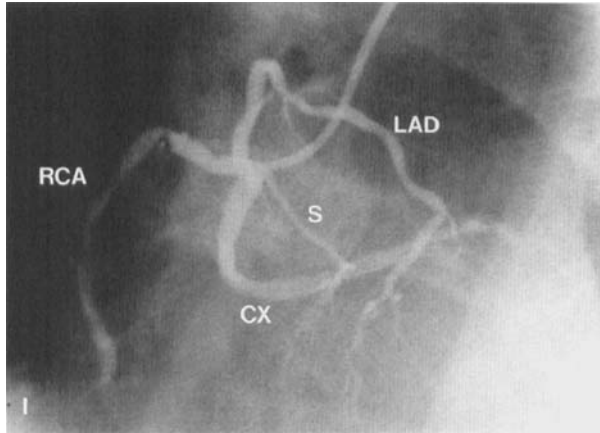


8G,H) (43). An RAO ventriculogram or aortogram will place the LMT on end, giving the appearance of a "dot" on the posterior aspect of the aorta (44). Effort syncope and myocardial infarction have been reported in one patient (53). A fifth subtype is a combination of "anterior," "septal," and "posterior" subtypes (Fig. 8I,J).

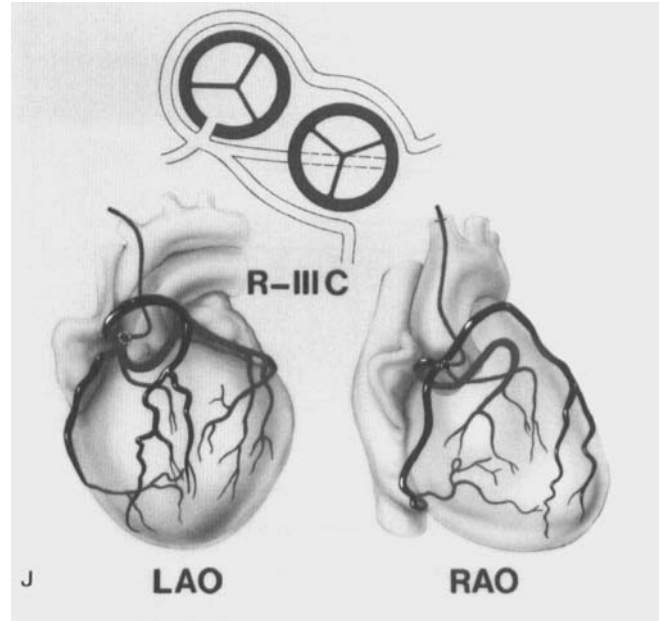
It is, at times, difficult to distinguish the five anatomic variants precisely by angiography. However, the course of initial loops demonstrated in the RAO (and LAO) projections, and the "dot" and "eye" method are help-

ful in identifying the correct anatomical course (43,44). Exercise testing with myocardial perfusion scanning may identify patients at risk for a future cardiac event, although a normal exercise test does confirm immunity from a future complication.

Ectopic origin of right coronary artery from the left sinus of Valsalva. In 135 patients, an anomalous RCA originated from an orifice located anterior to the left main ostium in the left sinus of Valsalva and passed between the aorta and pulmonary artery before reaching



Figs. 8I–J. I,J: Ectopic origin of left coronary artery from right, “combined” type, LAO projection. The LAD passes anterior to the pulmonary artery, the septal branch (S) intramurally, and the CX posterior to the aorta. There is diffuse coronary atherosclerosis with severe narrowing of the mid-right coronary artery (RCA).



the right AV groove (Fig. 9). This anomaly is suspected when the RCA ostium is unable to be located in the right sinus of Valsalva and collateral vessels are absent. The ectopic right coronary artery is difficult to cannulate because of its slit-like orifice and odd angulation. Ostial occlusion due to aortic expansion during exercise may result in myocardial ischemia (54). Clinical sequelae are rare, although angina pectoris, myocardial infarction, ventricular tachycardia, syncope, and sudden death in the absence of coronary atherosclerosis have been reported. (3,51,54–57)

Only one patient (0.7%) had the RCA orifice posterior to the left main ostium and a course posterior to the aorta. This patient had angina pectoris due to severe atherosclerotic LAD narrowing. There was no evidence of inferior wall ischemia secondary to the anomalous right coronary on a preoperative stress test.

Single coronary artery. The various patterns of single coronary artery are difficult to understand. The Lipton classification scheme, although incomplete, is the most practical for angiographers (58). The anomalous coronary artery is first designated with “R” or “L” depending upon whether the ostium is located in the right or left sinus of Valsalva. It is then designated as group I, II, or III. Group I has an anatomical course of either a right or left coronary artery. Group II anomalies arise from the proximal part of the normal right or left coronary artery, and cross the base of the heart before assuming the normal position of the inherent coronary artery. Group III describes the anomaly where the LAD

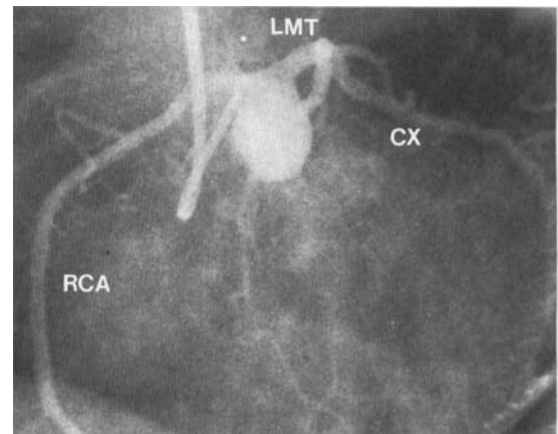


Fig. 9. Ectopic origin of right coronary artery (RCA) from left sinus of Valsalva, LAO projection. The anomalous right coronary artery passes between the pulmonary artery and aorta before assuming a normal position in the right AV groove. CX, circumflex; LMT, left main trunk.

and CX arise separately from the proximal part of the normal right coronary artery. The final designation refers to the relationship between the anomalous coronary artery and the aorta and pulmonary artery. The letters “A,” “B,” and “P” refer to “anterior,” “between,” and “posterior” patterns. We modified the Lipton’s classification by adding “septal” and “combined” types, designated as “S” and “C,” in order to describe the anatomical variations more precisely.

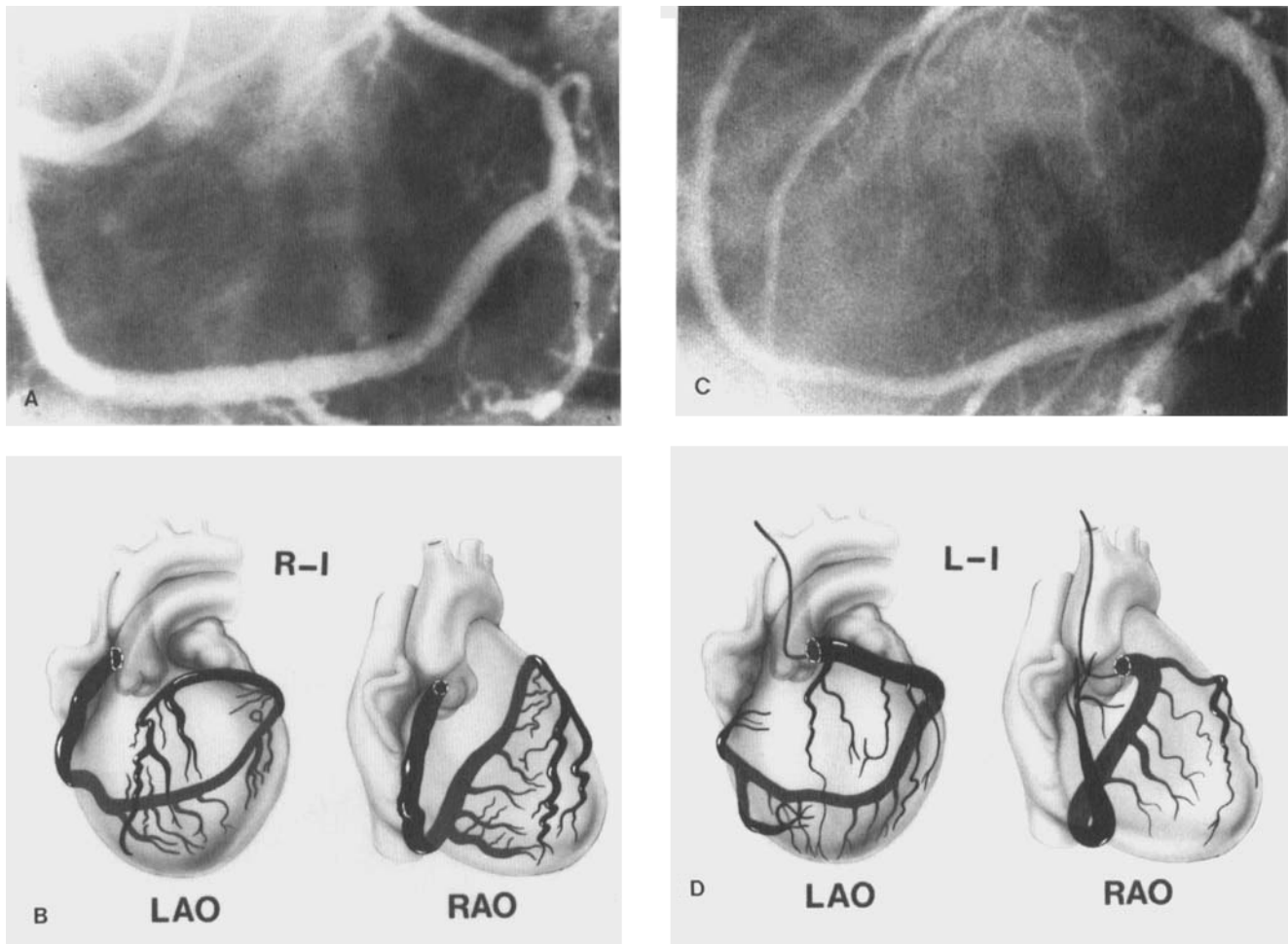


Fig. 10. A,B: Single coronary artery (R-I subtype). The right coronary artery perfuses the entire myocardium. The left coronary artery is congenitally absent. **C,D: Single coronary artery**

(L-I subtype). The markedly dominant left coronary artery perfuses the entire myocardium. The right coronary artery ostium is congenitally absent.

The R-I pattern refers to the rare instance where the RCA perfuses the entire heart (Fig. 10A,B). The L-I pattern occurs where the right coronary is congenitally absent and the CX is markedly dominant (Fig. 10C,D). The CX provides the posterior descending branch and ascends in the right AV groove where it provides branches to the right atrium and right ventricle. The extremely rare Group I anomalies generally have a benign clinical course (59), although a retired professional basketball player with an unsuspected R-I single coronary artery died suddenly during exercise (60).

In the type II single coronary artery, an anomalous coronary artery arises from the proximal part of the normal coronary. The R-II and R-III anomalies correspond to ectopic origin of the left coronary artery from the right sinus of Valsalva (Fig. 8A–J). The septal subtype is the most common pattern. In the L-II anomalies, the RCA arises from the proximal portion of the left coronary

artery (Fig. 11). In most instances, the anomalous RCA originates from the LMT and passes between the aorta and pulmonary artery before reaching the right AV groove. Rarely, the vessel passes anterior to the pulmonary artery or posterior to the aorta. No serious clinical sequelae were recognized in our small group of L-II variants.

Coronary artery fistulae. Coronary artery fistulae with large intracardiac shunts are rare in adults, since the majority are now detected and repaired during childhood. Fistulae draining into right heart chambers function as left to right shunts and may result in right ventricular volume overload. Patients with large fistulae may present with a continuous murmur, exertional dyspnea, effort intolerance, or congestive heart failure. Symptomatic patients with large fistulae should undergo surgical ligation of the fistulae at the drainage site. In this series, seven patients (3% of coronary artery fistulae)

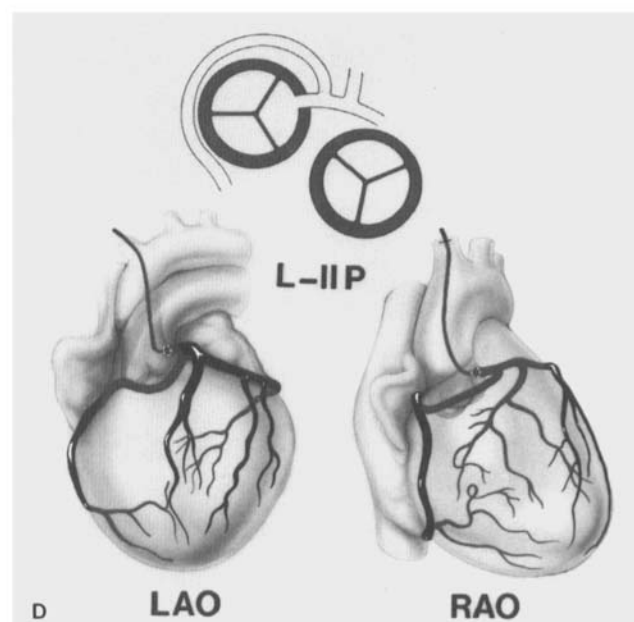
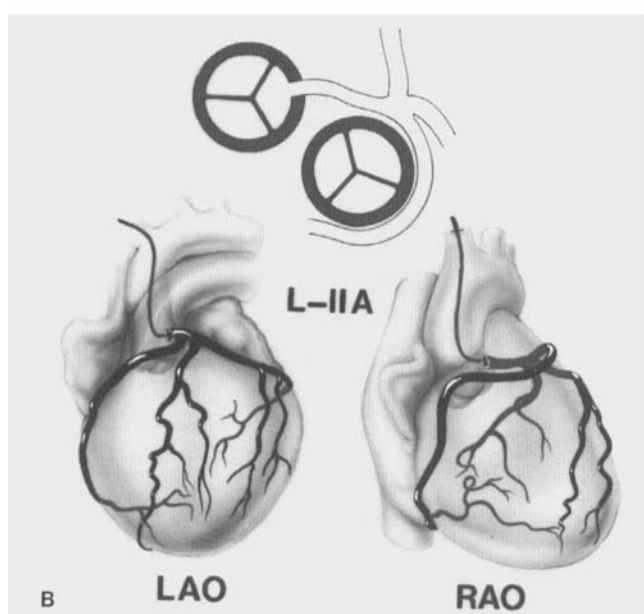
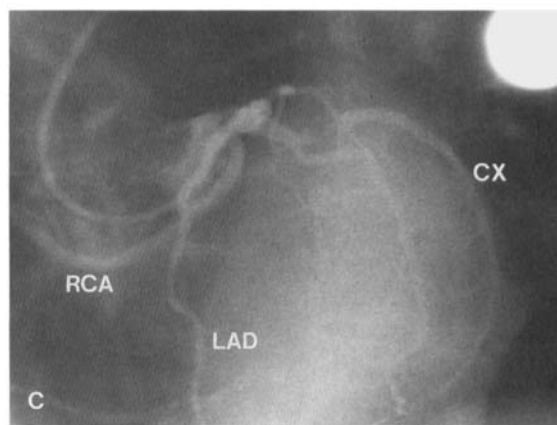
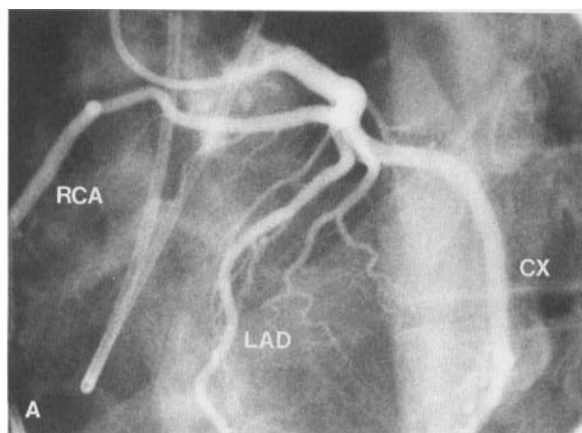


Fig. 11. A,B: Ectopic origin of right coronary artery (RCA) from proximal left coronary artery, LAO projection. The right coronary originates from the proximal part of left coronary artery and passes epicardially across the right ventricular outflow tract to the right AV groove. CX, circumflex; LAD, left anterior

descending artery. **C,D:** Ectopic origin of right coronary from left main trunk, LAO projection. The right coronary artery (RCA) originates from the left main trunk and courses posterior to the aorta. The CX has severe narrowing in its proximal portion. LAD, left anterior descending artery.

were operated solely to correct the huge fistulae. Those fistulae originated either from the RCA or the CX, and drained into one of the right cardiac chambers (Fig. 12).

SUMMARY

Isolated coronary anomalies occur in 1.3% of patients undergoing coronary arteriography. Eighty-seven percent are anomalies of origin or distribution, and 13% are coronary artery fistulae. Eighty-one percent of anomalies are considered to be benign and include: 1) separate origin of left anterior descending and left circumflex from

the left sinus of Valsalva; 2) ectopic origin of the circumflex from the right sinus of Valsalva; 3) ectopic coronary origin from the posterior sinus of Valsalva; 4) ectopic coronary origin from the posterior sinus of Valsalva; 5) ectopic coronary origin from the ascending aorta; 6) absent circumflex; 7) intercoronary communication; and 8) small size coronary artery fistulae.

Potentially serious anomalies constitute 19% of the group and include: 1) ectopic coronary origin from the pulmonary artery; 2) ectopic coronary origin from the opposite aortic sinus; 3) single coronary artery; and 4) large coronary artery fistulae. Most coronary artery

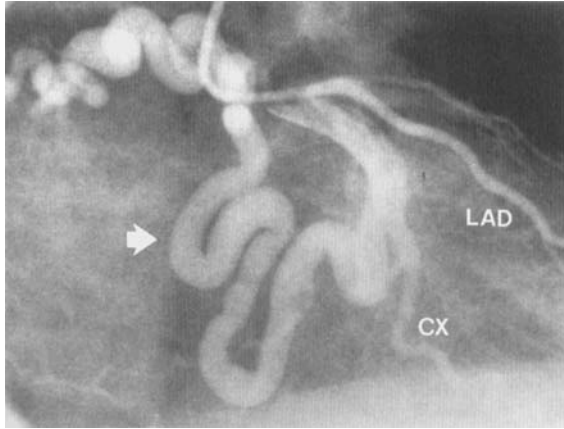


Fig. 12. Large coronary fistula (arrow) arising from the circumflex (CX) and draining into the right atrium, RAO projection. LAD, left anterior descending artery.

anomalies do not result in signs, symptoms, or complications and usually are discovered as incidental findings at the time of catheterization. Coronary artery anomalies require accurate recognition in order to ensure appropriate management.

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