Congenital variations of coronary artery anatomy

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During the past 20 years (1958 to 1978), more than 64,000 coronary arteriograms were performed at the Cleveland Clinic. During the course of this experience, a variety of anatomical variants were observed, some of which were clinically significant, causing symptoms, affecting cardiac function, or influencing prognosis. This report will attempt to review the spectrum of anomalies encountered during the first generation of coronary arteriography, define their incidences during a specific segment of time, and discuss their clinical significance.

Methods

During the first generation of coronary arteriography, many of the more unusual anomalies of the coronary circulation have been collected individually by staff members of the Department of Cardiology. An attempt has been made to compile these records, which include most, but not all, of the clinically significant anomalies. In 1972, a computerized registry was initiated that would serve as a repository of data for cardiovascular patients, initially those undergoing cardiac catheterization or surgical treatment at the Cleveland Clinic. Currently, the Cardiovascular Information Registry contains information on more than 42,000 patients. Data acquisition is more than 99% complete for all

patients undergoing catheterization or surgery from 1973 to 1978, and more than 98% complete for 1972 patients. This data base was surveyed for coronary anomalies identified during the first 7 years of the registry through 1978. Catheterization reports, clinical records, and in many instances cineangiograms were reviewed of patients with coronary anomalies for accuracy and completeness. Patients catheterized in other institutions were excluded. A coronary anomaly was defined as any abnormality of origin or distribution of the coronary arteries (but not including patients with dominance of the left coronary system or balanced dominance of the right and left coronary distributions). Six hundred one patients were identified with coronary anomalies (*Table 1*), representing 1.6% of all coronary arteriographic studies performed during these years (38,703). The preponderant indication for study of these patients was suspected arteriosclerotic heart disease. Thirteen percent had associated congenital heart lesions, and 4% had associated rheumatic heart disease. Seventy-nine percent were male and 21% female.

The most common anomaly encountered was separate origin of the anterior descending and circumflex arteries from the left anterior sinus of Valsalva or absence of a left main coronary artery. This congenitally short left main artery is associated with dominance of the left coronary distribution, and frequently, anomalies of the aortic valve. Ectopic origin of the circumflex division of the left coronary artery from the right coronary artery or sinus accounted for 29.1% of the anomalies, and an incidence of 0.45% in the population studied. Invariably the circumflex vessel arose from an orifice adjacent to the right coronary artery and passed to the

Table 1. Congenital variations of coronary artery anatomy; Cleveland Clinic Cardiovascular Information Registry (1972 to 1978 inclusive)

	No.	Incidence, %	Anomalies, %
Coronary arteriograms	38703	...	...
Total coronary anomalies	601	1.6	100
Absent LMT (separate origin of LAD and LCx)	185	0.48	30.8
Absent LCx (markedly dominant RCA)	3	0.008	0.5
Absent RCA (markedly dominant LCA)	8	0.02	1.3
Ectopic origin from aorta above sinuses of Valsalva	76	0.20	12.6
Right	68	0.18	11.3
Left	8	0.02	1.3
Ectopic origin LCx from right anterior sinus	176	0.45	29.2
Ectopic origin RCA from left anterior sinus	50	0.13	8.3
Ectopic origin LAD from RCA	13	0.03	2.2
Ectopic origin LCA from right sinus	9	0.02	1.5
Ectopic origin LAD and Cx (separately) from right sinus	4	0.01	0.7
Anomalous origin from MPA			
Right	1	0.003	0.17
Left (Bland-Garland-White Syndrome)	1	0.003	0.17
Other	6	0.02	1.0
Coronary artery fistula	78	0.20	13.0

LMT = left main coronary artery, LAD = left anterior descending artery, LCx = left circumflex artery, RCA = right coronary artery, LCA = left coronary artery, Cx = circumflex artery, MPA = main pulmonary artery.

right and posterior to the aorta to the left atrioventricular groove at which point its branches were distributed normally to the lateral and posterolateral aspects of the left ventricle. The anomalous vessel occasionally showed luminal irregularities in patients with other evidence of atherosclerotic disease.

Perhaps of greater clinical significance are the anomalous origins of the right or left coronary arteries from the opposite (left or right) sinuses of Valsalva. Fifty instances of ectopic origin of the right coronary artery from the left coronary sinus or left coronary artery were encountered constituting 8.3% of the anomalies (0.13% of the arteriographic series). Right coronary arteries that arose from the left coronary sinus invariably coursed between the aorta and pulmonary artery toward the right atrioventricular groove; ectopic origin of the right coronary artery from the anterior descending vessel was rare, with the anomalous vessel passing across the right ventricular outflow tract toward the right atrioventricular sulcus. In nine instances, the main trunk of the left coronary artery arose ectopically from the orifice adjacent to the right coronary artery in the left anterior sinus of Valsalva (or the anterior sinus of a bicuspid valve in one case), and in most instances the anomalous vessel appeared to course anteriorly and to the left, between the pulmonary outflow tract and the aorta. This accounted for 1.5% of the anomalies (0.02% of patients). A total of 17 patients with this type of anomaly have been collected in the accumulated experience to date. In four patients, the anterior descending and circumflex branches arose separately from the right anterior sinus of Valsalva (0.7% of anomalies, 0.01% of arteriograms). The anomalous circumflex vessel coursed posteriorly behind the root of the aorta, and the anterior descending artery was directed anteriorly, either anterior to the pulmonary artery or between the pulmonary artery and the aorta. Thirteen instances of ectopic origin of the anterior descending artery from the right coronary artery or sinus were noted (2.2% of anomalies, 0.03% of the angiographic series). In most instances the anomalous anterior descending vessel appeared to course anterior to the pulmonary outflow tract en route to the anterior interventricular sulcus.

Only one example of anomalous origin of the left coronary artery from the main pulmonary artery was found during the 7-year study period (0.15% of anomalies), but in the past 20 years, 11 such patients have been encountered, all but one of whom had the intermediate or adult form of the anomaly with left-to-right shunting into the pulmonary artery. One patient has been observed with anomalous origin of only the circumflex division of the left coronary artery from the main pulmonary artery, and one with anomalous origin of the right coronary artery from the pulmonary artery.

Fistulous communications of coronary arteries with cardiac chambers or the superior vena cava occurred in 78 patients and constituted 14% of the anomalies (0.2% of the angiographic series). Frequently, multiple fistulas of different origins drained into a common site. In 29 patients a fistulous communication between the anterior descending artery or left main coronary artery and the pulmonary artery was noted. Such fistulas are usually small, and the magnitude of the shunt is inconsequential, but rare instances of giant fistulas with sizable shunting have been encountered in the 20-year experience. (*Table 2*).

At the present time, only three cate-

Table 2. Coronary artery fistulas

Origin	Recipient SVL/RA	RV	PA	LA	LV	Total
Single						
RCA	2	2	4	2	1	11
LCx	0	0	5	2	4	11
LAD	0	1	29	0	9	39
Total	2	3	38	4	14	61
Multiple	1	1	10	2	3	17
Total	3	4	48	6	17	78

SVC/RA = superior vena cava or right atrium, RV = right ventricle, PA = pulmonary artery, LA = left atrium, LV = left ventricle, and other abbreviations as in *Table 1.*

gories of coronary anomalies are of clinical importance and negative therapeutic consideration: ectopic origin of the left coronary artery passing between the pulmonary artery and the aorta, anomalous origin of the left coronary artery from the pulmonary artery, and large coronary fistulas. Ectopic origin of the left coronary artery from the right anterior sinus of Valsalva, when it courses between the pulmonary artery and the aorta, has been associated with sudden death during strenuous exertion. It has been hypothesized that myocardial perfusion is compromised by diminished flow through the anomalous vessel, either by compression from rising pressure within the two great vessels, or flattening of its lumen from angulation at its origin as it passes acutely anteriorly and to the left around the anterior margin of the aorta as it courses toward the anterior interventricular sulcus. Among ten such patients we have observed (including one studied elsewhere), four have undergone surgical treatment with bypass grafts to relieve myocardial ischemia.

Anomalous origin of the left coronary artery from the pulmonary artery is a well-known clinical entity. In its infantile form, without left-to-right shunting of the anomalous left coronary artery into the pulmonary artery with intercoronary collateral, this anomaly is uniformly fatal during the first year or two of life. This is associated with early development of heart failure, severe mitral insufficiency with an underdeveloped mitral valve apparatus, and frequently a pattern of myocardial infarction on the electrocardiogram due to ischemic fibrosis of left ventricular myocardium. Urgent surgical intervention is indicated, as antegrade flow into the anomalous left coronary artery can be established with a venous autograft, or direct reimplantation of the anomalous vessel into the aorta. In the transitional or adult form, left-to-right shunting occurs through intercoronary collateral communications with the anomalous left coronary vessel draining into the pulmonary artery. This may or may not be associated with mitral insufficiency. Although the natural history of this complex has not been well defined, atrial fibrillation, congestive heart failure, and sudden death have been recorded. Surgical correction can be performed electively, with venous bypass grafts and ligation of the anomalous vessel or direct anastomosis of the anomalous vessel with the aorta.

Most coronary arteriovenous fistulas are small and do not require surgical treatment. Isolated reports of sudden death, rupture, or bacterial infection of large fistulas suggest that giant fistulas or those associated with large left-to-right shunts should be treated surgically with simple ligation.

References

1. Chaitman BR, Lespérance J, Saltiel J, et al: Clinical, angiographic, and hemodynamic findings in patients with anomalous origin of

the coronary arteries. Circulation **53:** 122–131, 1976.
2. Levin DC, Fellows KE, Abrams HL: Hemodynamically significant primary anomalies of the coronary arteries; angiographic aspects. Circulation **58:** 25–33, 1978.
3. Liberthson RR, Dinsmore RE, Bharati S, et al: Aberrant coronary artery origin from the aorta; diagnosis and clinical significance. Circulation **50:** 774–779, 1974.
4. Cheitlin MD, DeCastro CM, McAllister HA: Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva; a not-so-minor congenital anomaly. Circulation **50:** 780–787, 1974.
5. Chaitman BR, Bourassa MG, Lespérance J, et al: Anomalous left coronary artery from pulmonary artery; an eight year angiographic follow-up after saphenous vein bypass graft. Circulation **51:** 552–560, 1975.
6. Askenazi J, Nadas AS: Anomalous left coronary artery originating from the pulmonary artery; report on 15 cases. Circulation **51:** 976–987, 1975.
7. Effler DB, Sheldon WC, Turner JJ, et al: Coronary arteriovenous fistulas; diagnosis and surgical management; report of fifteen cases. Surgery **61:** 41–50 1967.