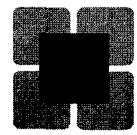

# Congenital heart diseases associated with coronary artery anomalies

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■ Coronary artery anomalies are commonly associated with congenital heart disease. It is important to recognize these anomalies to avoid errors in diagnosis, decrease technical difficulties in angiography, and prevent inadvertent injury in surgery. We found that of 66,884 patients who underwent coronary arteriography between 1972 and 1982 at the Cleveland Clinic Foundation, 1,000 had coronary anomalies. Of these, 101 had associated congenital anomalies: 29 had mitral valve prolapse, 18 had bicuspid aortic valves, 16 had tetralogy of Fallot, 11 had corrected transposition, 10 had a univentricular heart, 6 had coarctation of the aorta, 3 had ventricular septal defects, and 8 had miscellaneous congenital heart defects. The most common coronary anomaly was ectopic origin of a coronary artery: 30 from the sinus of Valsalva, 12 from the ascending aorta, 11 from the pulmonary artery. Nineteen patients had no left main trunk. Thirteen patients had coronary artery fistulas and 21 had miscellaneous coronary anomalies.

□INDEX TERMS: CORONARY VESSEL ANOMALIES; HEART DEFECTS, CONGENITAL □CLEVE CLIN J MED 1990;57:147–152

CORONARY ARTERY ANOMALIES are not rare in congenital heart disease.[1] Certain coronary anomalies are difficult to diagnose in complex forms of congenital heart disease, but a high index of suspicion for coronary anomalies in certain congenital heart diseases may decrease the technical difficulties the angiographer faces. Most important, recognizing the variations in origin and distribution of the coronary arteries may help avoid inadvertent injuries to coronary arteries and coronary circulation during surgery for complex congenital heart disease. This study describes the association of congenital heart disease with coronary anomalies.

■ *See related editorial by Moller (pp 178–180)*

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## MATERIALS AND METHODS

Our patient population consisted of all patients undergoing coronary arteriography at the Cleveland Clinic Foundation from 1972 through 1982. Coronary arteriographic findings were stored in a computerized registry. Hobbs et al reported their findings in 1,000 patients with coronary artery anomalies among the consecutive 66,884 patients who underwent coronary arteriography at the Cleveland Clinic Foundation.[2] The incidence of congenital heart disease and the patterns of coronary anomalies were analyzed in the 1,000 patients.

## RESULTS

Of the 1,000 patients with coronary artery anomalies, 101 had congenital heart disease. In 14 of these patients,

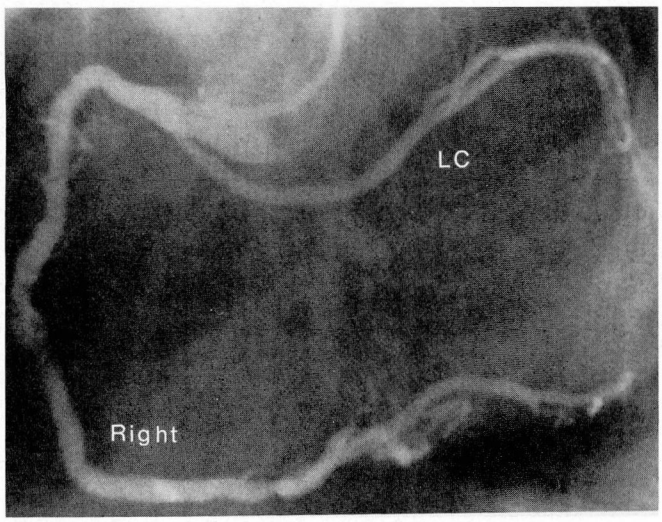


FIGURE 1. Anomalous origin of the left circumflex artery from the right sinus of Valsalva in a patient with mitral valve prolapse (left anterior oblique projection). *LC* = left circumflex coronary artery.

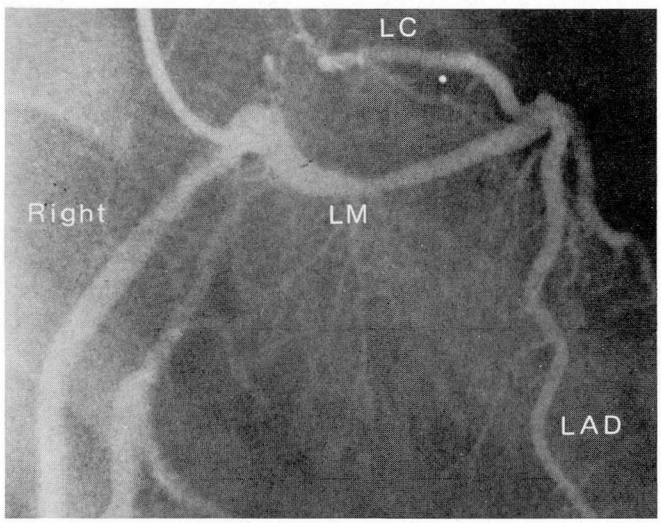


FIGURE 2. Left coronary artery originating from the right coronary artery in a patient with mitral valve prolapse (left anterior oblique projection). *LAD* = left anterior descending coronary artery; *LC* = left circumflex coronary artery; *LM* = left main trunk coronary artery.

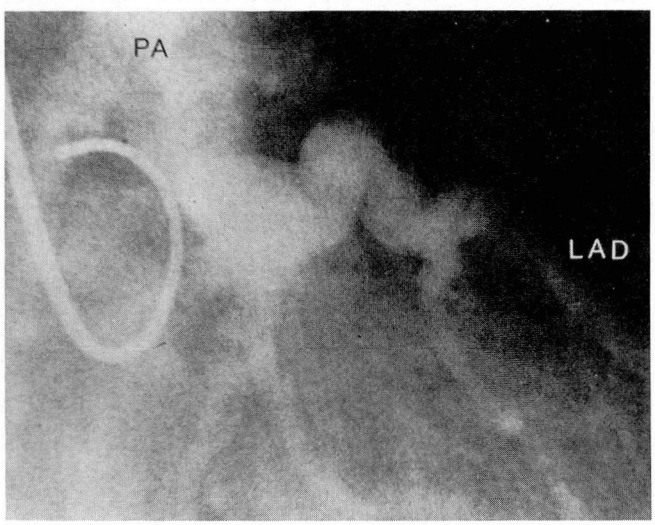


FIGURE 3. Fistula from the left anterior descending coronary artery to the pulmonary artery (right anterior oblique projection). *LAD* = left anterior descending coronary artery; *PA* = pulmonary artery.

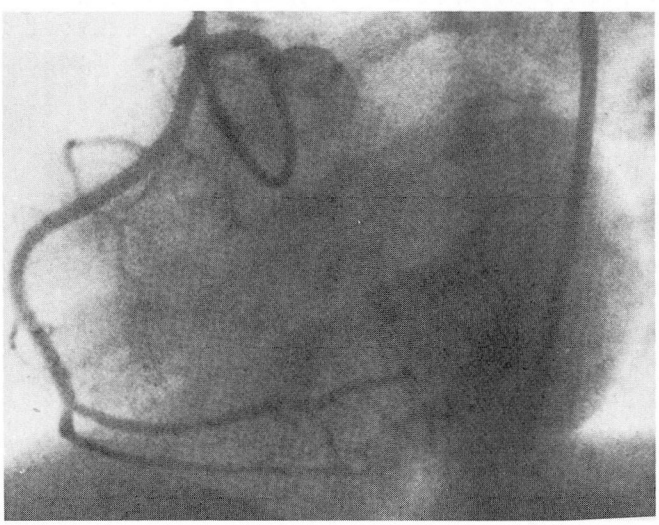

FIGURE 4. Anomalous origin of the right coronary artery from the left sinus of Valsalva in a patient with tetralogy of Fallot (left anterior oblique projection).

the affected coronary arteries could not be visualized because of technical difficulties. The congenital heart defects associated with coronary artery anomalies are as follows: 29 patients had mitral valve prolapse, 18 had bicuspid aortic valves, 16 had tetralogy of Fallot, 11 had corrected transposition, 10 had a univentricular heart, 6 had coarctation of the aorta, 3 had ventricular septal defects, and 8 had miscellaneous congenital heart defects.

In the 29 patients with mitral valve prolapse (mean age 58 ± 8 years; range 45 to 71 years), no findings sug-

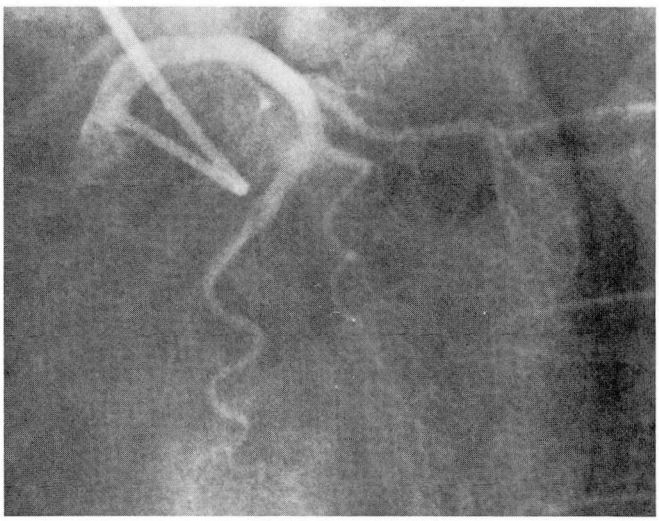

FIGURE 5. Left anterior descending artery originating from right sinus of Valsalva in a patient with tetralogy of Fallot (left anterior oblique projection).

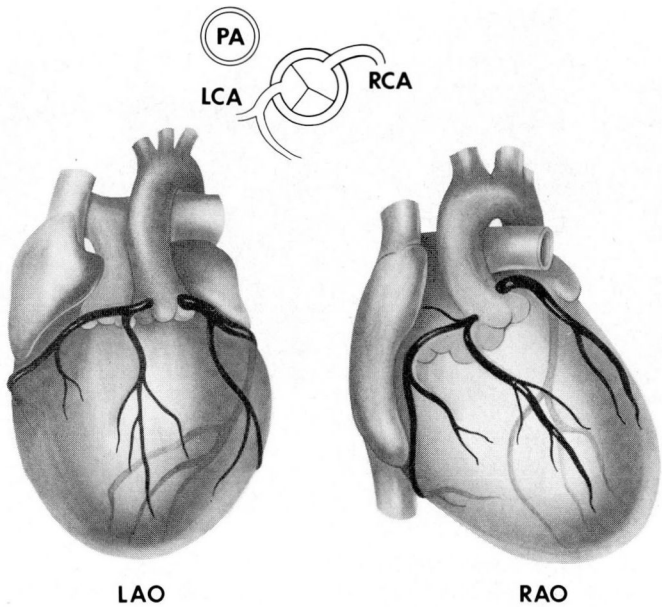


FIGURE 6. Diagrammatic representation of coronary artery anatomy in corrected transposition of the great vessels. The morphological right coronary artery is a large and dominant vessel that arises from the left sinus of Valsalva and supplies the left-sided morphological right ventricle. The morphological left coronary artery arises from the right sinus of Valsalva and supplies the right-sided morphological left ventricle. The noncoronary sinus is located anteriorly. (*LAO* = left anterior oblique projection; *LCA* = left coronary artery; *PA* = pulmonary artery; *RAO* = right anterior oblique projection; *RCA* = right coronary artery.)

gested acquired mitral valve disease. In this group, the left circumflex artery originated from the right coronary artery in five patients and from the right sinus of Valsalva in six patients (*Figure 1*). In eight patients, the left anterior descending and left circumflex arteries originated from separate orifices in the left sinus of Valsalva. The left coronary artery originated from the ascending aorta above the left sinus in one patient and from the right coronary artery in another patient (*Figure 2*). In four patients, the right coronary artery had an anomalous origin; one from the left coronary artery, one from the left sinus of Valsalva, one from the noncoronary sinus, and one from the ascending aorta. In four patients, there were fistulous communications: three had fistulas from the left anterior descending artery and one had a fistula from the right coronary and circumflex arteries to the pulmonary artery.

Eighteen patients had a bicuspid aortic valve (mean age 55 ± 11 years; range 34 to 82 years). In seven patients the left anterior descending and left circumflex arteries had separate origins in the left sinus of Valsalva. In five patients, the left circumflex artery had an anomalous origin in the right sinus of Valsalva. In five patients, the right coronary artery had an anomalous origin: the origin was from the ascending aorta in three patients, from the left sinus of Valsalva in one, and from the left circumflex artery in one. In one patient, there was fistulous communication between the left coronary artery and the pulmonary artery (*Figure 3*).

There were 16 patients with tetralogy of Fallot (mean age 19 ± 12 years; range 7 to 42 years). An anomalous origin of the right coronary artery was the most common anomaly (11 patients): from left sinus of Valsalva in four patients (*Figure 4*), from left coronary artery in three, from the ascending aorta in three, and from the pulmonary artery in one. The left circumflex and left anterior descending arteries had an anomalous origin from the right sinus of Valsalva in two patients (*Figure 5*). Three patients had fistulas: two had fistulas from the right coronary artery to the right ventricle and one had multiple fistulas, including a fistula from the left anterior descending artery to left ventricle, from the left circumflex artery to left ventricle, and from the right coronary artery to the right ventricle.

There were 11 patients with corrected transposition of the great vessels (mean age 46 years; range 25 to 67 years). The coronary artery anatomy was consistent in all cases (*Figure 6*). The morphological right coronary artery arose

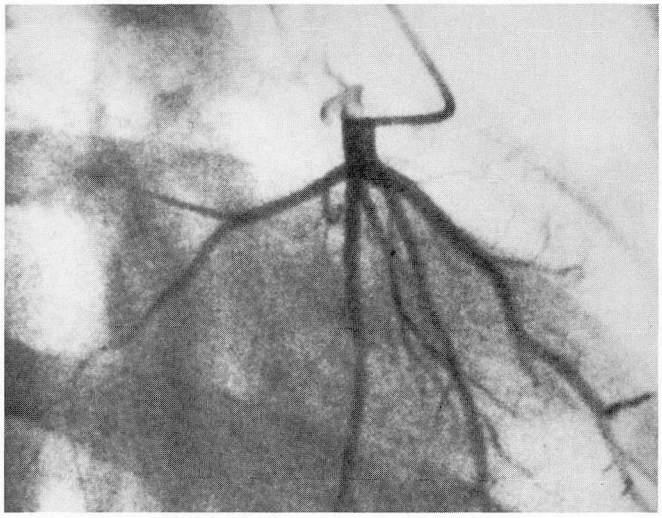

FIGURE 7. The morphological left coronary artery arising from the right sinus of Valsalva and supplying the right-sided morphological left ventricle (right anterior oblique projection).

from the left sinus of Valsalva or just above it and supplied the left-sided morphological right ventricle. In all cases, the morphological right coronary system was dominant. The morphological left coronary artery arose from the right sinus of Valsalva or just above it and supplied the right-sided morphological left ventricle (*Figure 7*).

Ten patients had a univentricular heart (mean age 18 years; range 3 to 38 years). In seven patients, the right coronary artery was the dominant vessel. Large branches originating from the right coronary artery with parallel distribution supplied the anterior and lateral wall of the heart. A large branch originating from the right coronary artery had a descending course similar to the left anterior descending artery. This large branch acted as a determining artery when an outlet chamber was present (*Figure 8*).

Six patients had coarctation of the aorta (mean age 50 ± 30 years; range 31 to 61 years). The left anterior descending and left circumflex arteries had separate origins from the left sinus of Valsalva in three of six patients. In one patient, the circumflex artery originated from the right coronary artery. In one patient, the left coronary artery originated from the ascending aorta above the left sinus of Valsalva. This same patient was also noted to have a fistula from the right coronary artery to the superior vena cava. In another patient in this group, the right coronary artery originated from the ascending aorta above the junction of the left and right sinuses of Valsalva.

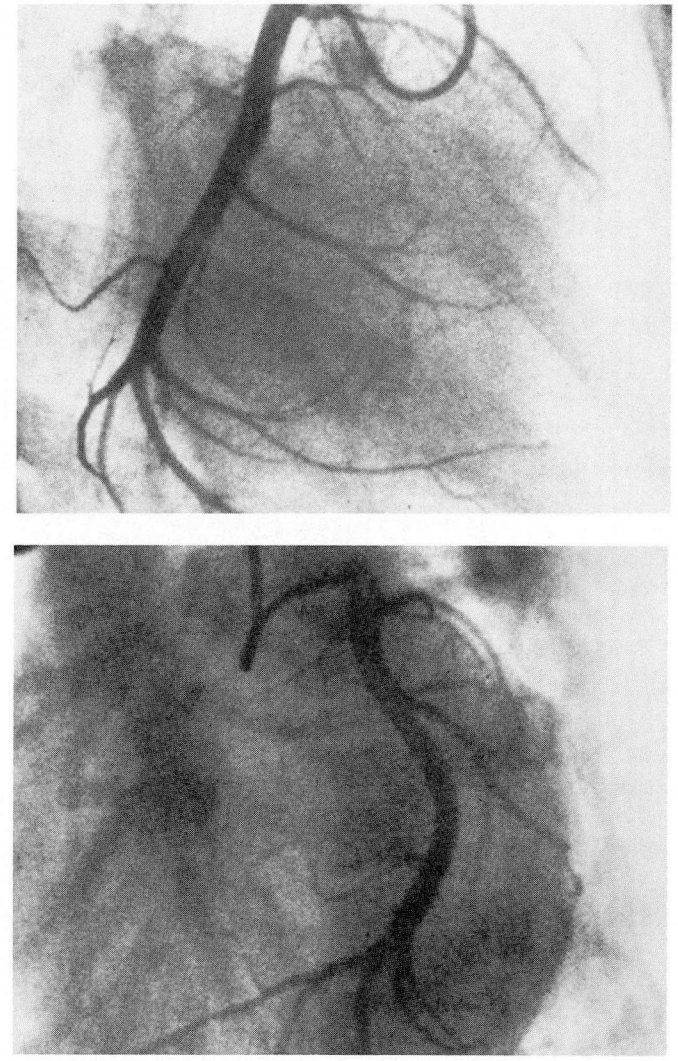


FIGURE 8. The dominant right coronary artery in a patient with a univentricular heart with an outlet chamber of the aorta anterior and to the left in (A) right anterior oblique and (B) left anterior oblique projections. The right coronary artery arises posteriorly and supplies the anterolateral portion of the common ventricle in a fan-like pattern.

Three patients had a ventricular septal defect (mean age 25 ± 20 years; range 4 to 28 years). One patient had the circumflex artery originating from the right coronary artery and coursing posterior to the aorta, another had the left coronary artery originating from the ascending aorta above the left coronary sinus, and the third had a fistula from the left coronary artery to the left ventricle.

Various coronary anomalies were associated with miscellaneous congenital heart defects in eight patients.

Two patients had pulmonary stenosis, two had atrial septal defects, two had complete transposition of the great vessels, one had cor triatriatum, and one had a double-outlet right ventricle. Six patients had anomalous origins of coronary arteries, two patients had left anterior descending and left circumflex coronary arteries that had separate origins in the left sinus of Valsalva, and one patient had a coronary fistula.

## DISCUSSION

Our study group is selected from a larger group of patients with coronary artery anomalies. This point should be considered in interpreting results. Unlike several previous studies, the results of this study do not represent the true incidence of various types of coronary anomalies in certain congenital heart diseases because not all cases of congenital heart disease evaluated in the same period were analyzed for coronary anomalies.

A high incidence of an absent left main trunk in patients with bicuspid aortic valve has been reported by other investigators.[3] Autopsy reports have shown that bicuspid aortic valve is frequently associated with a short or absent left main trunk. Our findings confirm these reports. Seven of 18 patients with bicuspid aortic valve had separate origins of the left anterior descending and circumflex arteries in the left sinus of Valsalva.

These anomalies, which may stem from a developmental anomaly of the aortic root, have important clinical implications. When patients with a bicuspid aortic valve undergo diagnostic catheterization, the separate coronary orifices of the left anterior descending and circumflex arteries may cause technical problems. When coronary artery perfusion is used for myocardial preservation in these patients during open heart surgery, cannulation of one left coronary orifice may lead to inadequate perfusion of the myocardium supplied by the other left coronary artery branch if that is not also cannulated. The left coronary artery provides the dominant circulation in the majority of patients with a bicuspid aortic valve.[4] This left coronary dominance makes attention to these technical problems all the more important in patients with a bicuspid aortic valve.

The left main trunk was absent in 9 of 18 patients with coarctation of the aorta and coronary artery anomalies. Although none of these patients had a bicuspid aortic valve, the high overall incidence of bicuspid aortic valve [3] in this congenital anomaly suggests that a common developmental anomaly may underlie this congenital heart disease.

Mitral valve prolapse was the most common congenital anomaly in the 1,000 cases we analyzed. More than one-third of our patients with mitral valve prolapse and coronary anomalies had an anomalous origin of the left circumflex artery, either from the right coronary artery or from the right sinus of Valsalva. This is a very common coronary anomaly in the general population as well.[2] Three patients had an anomalous right coronary artery originating from the left sinus of Valsalva, left coronary artery, or noncoronary sinus. Should such a patient need valve replacement, the cardiac surgeon should be warned about the anomalous coronary artery to avoid accidentally interrupting the vessel during surgery.

In patients with tetralogy of Fallot, different incidences of anomalous coronary artery distribution have been reported. Most angiographic studies show coronary anomalies in 5% to 10% of cases.[5,6] The left anterior descending artery originating from the right coronary artery was reported as the most common artery anomaly associated with tetralogy of Fallot.[5]

In our study, however, 69% of patients with coronary anomalies and tetralogy of Fallot had anomalous origin of the right rather than the left coronary artery. The single most common coronary anomaly was the origin of the right coronary artery from the left sinus of Valsalva, which was present in four patients. In three patients, the right coronary artery originated from the left coronary artery and, in three others, from the ascending aorta. Only one patient had an anomalous origin of the left anterior descending from the right sinus of Valsalva. Most of these abnormal patterns of distribution, in themselves, do not lead to recognizable clinical problems. However, when corrective surgery is attempted, increased morbidity and mortality may occur.[7] The right coronary artery with anomalous origin from the left sinus or left coronary artery generally courses over the right ventricular outflow tract. This course may lead to inadvertent transection of the coronary artery during surgery. It is very important to define the precise course of the anomalous coronary artery preoperatively in tetralogy of Fallot.

Our findings in patients with corrected transposition parallel previous reports.[8] A consistent coronary arterial pattern in all patients was helpful in identifying the complex cardiac anatomy. The morphological right coronary artery was dominant in all of these patients. The precise definition of the course and origin of the coronary arteries preoperatively is important in preventing inadvertent injury to the coronary arteries during surgery to correct this complex congenital heart disease.

Reports on coronary artery pattern vary according to the type of univentricular heart.[9,10] Changes at the origin and distribution of coronary arteries are very common in the univentricular heart. In most of the cases, the coronary artery branches demarcate the outflow tract. The location of the proximal segments and course of the coronary arteries can present major surgical problems in patients with infundibular pulmonic stenosis.

The incidence of coronary anomalies in congenital heart disease is high. Knowledge of their various patterns and occurrence assist in diagnosis and treatment. Diagnosing the coronary anomaly before surgical treatment may prevent morbidity and mortality.

ACKNOWLEDGMENT

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