A Later Stage of Anomalous Coronary Circulation with Origin of the Left Coronary Artery from the Pulmonary Artery

Coronary Artery Steal

By Arthur E. Baue, M.D., Stanley Baum, M.D., William S. Blakemore, M.D., and Harry F. Zinsser, M.D.

SUMMARY
A 45-year-old woman with angina pectoris, beginning at the age of 16 and progressing to produce incapacitation, was found to have an anomalous left coronary artery arising from the pulmonary artery. Because of the severity of symptoms and progression of disability, the anomalous artery was ligated at its origin from the pulmonary artery. Pressure in the left coronary system increased from 35/10 to 105/30 mm Hg. Since operation the patient has been relieved of her symptoms. There was no evidence of increased myocardial ischemia after ligation. This indicates that operative treatment of this anomaly can be carried out successfully in the older age group. It is also suggested that a fourth stage of progression of this anomaly occurs in which there is an exaggeration of the intercoronary communications between the right and left coronary systems. This produces such a large arteriovenous shunt that blood flow to the myocardium is again reduced, resulting in a "coronary steal syndrome."

Additional Indexing Words:
Ligation, coronary artery Angiography Angina pectoris
Mitral incompetence Arteriovenous fistula

The most common congenital anomaly of the coronary circulation is a left coronary artery arising from the pulmonary artery. Many patients with this problem have died in infancy, but a number of individuals have survived beyond the early critical period into adult life. This led to descriptions of two separate, fixed forms of the anomaly:1-3 (1) the infant type, in which satisfactory intercoronary anastomoses between the right coronary artery and the anomalous left coronary artery did not develop, and (2) the adult type, in which abundant anastomoses developed, permitting adequate blood supply to the left coronary distribution and prolonged survival. More recently, Edwards4 suggested that this concept of fixed functional types be abandoned, and he, along with Nadas and associates5 and Talner and associates,6 described three progressive phases of this abnormality. In the initial neonatal phase there is high pressure perfusion of the anomalous artery from the pulmonary artery so that the infant appears normal at birth and for a short time afterward. The second phase is a critical transition period in which pulmonary artery pressure decreases and survival depends upon development of adequate intercoronary vessels. At this time, evidence of distress and myocardial ischemia may be present. In the third phase, the presence of a rich collateral circulation permits survival into adult life.
Previous ischemic changes determined by electrocardiography have been found to improve. However, prior cardiac damage from ischemia and fibrosis during the transition period may result in dilatation of the left side of the heart and secondary mitral insufficiency. This may become a significant and severe problem. Some individuals have survived the critical transitional period only to die suddenly in adult life, presumably from acute arrhythmia. Coronary angiography and observations made during surgical exposure of the anomalous vessel provided evidence that blood in the left coronary artery flowed in a retrograde direction into the pulmonary artery. It was suggested that the proper treatment would be ligation of the anomalous artery at its origin. This procedure has been done in a number of patients and has been satisfactory in infants and children in whom adequate collateral circulation between the two coronary arteries has been demonstrated. However, infants without adequate collateral communications have failed to survive ligation. Therefore, in infants, surgical treatment by ligation is limited to those who have demonstrable significant intercoronary communications. If such communications are not sufficient, treatment by a vascular graft from the aorta to the left coronary artery may be necessary.

The oldest patient reported to survive operative treatment by ligation was a 16-year-old girl. This patient was asymptomatic before operation, but postoperatively developed persistent electrocardiographic changes indicative of myocardial ischemia. Presumably, the collateral communications were inadequate even though she had survived to the age of 16 without symptoms. Recently, the case of a 29-year-old patient with this anomaly who had no symptoms but had electrocardiographic findings of an anterolateral myocardial infarction was reported. Because of the difficulties in the previously reported 16-year-old patient treated by ligation, surgical treatment was not recommended for this patient in spite of the recognition that acute arrhythmias seem to be a frequent cause of sudden death in these individuals. Thus, the treatment of such patients presents a dilemma at the present time. The purpose of the present communication is to report the successful treatment of a 45-year-old woman with this anomaly, in whom a fourth phase developed which could be called a "coronary steal syndrome."

Report of Case

A 45-year-old woman was admitted to the Graduate Hospital of the University of Pennsylvania on September 15, 1966, with a complaint of chest pain on exertion and an admitting diagnosis of rheumatic mitral insufficiency. During early childhood she had pneumonia on three occasions and frequent tonsillitis and joint pains. At the age of 10 she was found to have a heart murmur and was thought to have rheumatic heart disease. At the age of 16 she became pregnant. Interruption of the pregnancy was suggested because of her heart disease, but she was allowed to complete it. There was no difficulty in late pregnancy and she was delivered of a normal child. After the birth of this child, she had her first attack of severe crushing chest pain while chopping wood. The pain radiated to her neck and down her left arm. Later she developed dyspnea on exertion and palpitations and was thought to have congestive heart failure. Intermittent attacks of crushing chest pain with exertion continued. Six years before admission she was studied at another hospital and right cardiac catheterization findings were normal. A left-sided study resulted in injury to the femoral artery; this was repaired satisfactorily but the study was not completed. One year before admission there was a marked change, with more frequent attacks of angina brought on by simple tasks such as combing her hair. Along with the anginal pain, she complained on admission of decreased exercise tolerance, paroxysmal nocturnal dyspnea, and marked fatigue.

On examination she weighed 119 pounds. Blood pressure was 100/60 mm Hg in the right arm. There was a grade II-III somewhat musical systolic murmur at the apex and a prominent blowing systolic murmur with a less musical quality along the left sternal border at the third and fourth interspaces. No diastolic or continuous murmurs were heard. The second heart sound at the pulmonic region was within top limits of intensity. The right dorsalis pedis pulse was diminished. There were no other abnormalities on physical examination.

Fluoroscopy and chest x-ray showed the heart to be moderately enlarged, with a so-called "mi-
tral configuration." The peripheral vessels were of top normal prominence. The lung fields were clear. The electrocardiogram suggested left ventricular hypertrophy with possible superimposed localized damage in the anteroseptal region. The pattern in lead aVL showed a prominent Q wave and an inverted T wave compatible with a possible old infarct (fig. 1). From the clinical picture, with angina beginning at the age of 16, the diagnosis of an anomalous coronary artery was considered. Cineangioraphy with injection of contrast material into the aortic root showed an extremely large right coronary artery arising from the aorta (fig. 2A). No left coronary ostium was seen in the usual location. Later in this sequence, extensive dilated collateral vessels were seen arising from the right coronary artery and re-forming to enter the left coronary system, which emptied into the main pulmonary artery (fig. 2B, C, and D). This was also seen in the left anterior oblique projection. The extent of the collateral supply between the right and left coronary systems was impressive. Right heart catheterization showed a pressure of 30/10 mm Hg in the pulmonary artery and 30/0 mm Hg in the right ventricle. The right atrial pressure was normal. The pressure in the aorta was 130/80 mm Hg and in the left ventricle, 130/0 mm Hg.

Surgery was recommended because of the progression of her disability. She was operated upon on October 26, 1966, through an anterolateral thoracotomy incision in which the sternum was divided down to the third interspace and this interspace was opened on the left side. This provided excellent exposure of the pulmonary artery. An extremely large left coronary artery arising from the posterior aspect of the pulmonary artery was found. Within 2 cm it divided into a circumflex and an anterior descending branch. The vessel was extremely tortuous and thin-walled so that blood could be seen flowing

Figure 1
Preoperative electrocardiogram.
Figure 2

(A) Supravalvular thoracic aortogram in the right antero-oblique projection showing a large right coronary artery arising from the right anterior sinus of the aortic valve. No coronary artery is visualized arising from the left anterior sinus. (B) Three seconds after the injection of contrast material, beginning opacification is seen in the left coronary artery which has filled in a retrograde direction through large collateral channels communicating with the right coronary artery. (C) Diagrammatic representation of B. (D) Six seconds after the injection of contrast material, a film in the right antero-oblique projection demonstrates filling of the pulmonary artery from the left coronary artery.
through it. There was no evidence of coronary atherosclerosis. A marked thrill could be felt over this vessel. The segment of left coronary artery from the pulmonary artery to its bifurcation was mobilized. At this time, aortic pressure was 95/70 mm Hg, pulmonary artery pressure was 20/10 mm Hg, and pressure in the left coronary artery was 35/10 mm Hg. With occlusion of the left coronary artery at its origin, the pressure in this vessel rose to 105/30 mm Hg (fig. 3). The electrocardiogram did not change during 10 minutes of occlusion. Therefore, the anomalous artery was ligated close to the pulmonary artery, leaving the communication between the anterior descending and circumflex branches intact.

The patient did well for 24 hours after operation. She then developed frequent premature atrial contractions and occasional bursts of atrial tachycardia. Digitalization was begun, with 0.6 mg of digitoxin given. Later the rate increased to 160 per minute, with a sinus tachycardia. Intravenous infusion of lidocaine (Xylocaine: 250 mg in 500 cc of 5% dextrose in water) was begun. Inadvertently, 40 mEq of potassium chloride was added to the infusion, which was given rapidly. This was followed by cardiac standstill. External cardiac massage produced ventricular fibrillation, and external electrical defibrillation was quickly carried out. There were no sequelae from this episode. Infusion of Xylocaine in a small dose was continued for 24 hours. Over the next several days the patient's cardiac rhythm varied from atrial flutter to atrial fibrillation. After digitalization was completed her rate and rhythm became normal. She had an uneventful recuperation following this.

Postoperatively, the apical systolic murmur was less pronounced, and a faint pulmonic systolic murmur was still heard. She was discharged on the sixteenth postoperative day. An electrocardiogram at the time of discharge showed no significant change from that on admission. Serum isoenzyme studies on the third postoperative day, including lactate dehydrogenase, serum glutamic oxalacetic and pyruvic transaminase, were no higher than would be expected from thoracotomy alone.

Since discharge the patient has been completely asymptomatic. She has resumed full activity and has had no further episodes of chest pain or shortness of breath. On January 4, 1967, the apical systolic murmur was no longer heard. At 8 months after operation she has had an excellent result.

**Discussion**

The original reports of anomalous coronary arteries were based on autopsy studies by Brooks (1886), Abbott (1908), and Abrikossoff (1911). The clinical syndrome produced by these anomalies was first described in 1933 by Bland and associates. Since that time, anomalous origins of both the right and
the left coronary arteries and fistulae of the coronary arteries into various chambers of the heart, particularly the right atrium, have been described. The sinus of Valsalva, from which the left coronary artery arises, is in very close proximity to the septum of the truncus arteriosus. A slight displacement of its angle would result in location of the left coronary ostium within the pulmonary artery. This seems to be the reason why this anomaly is the most common of these problems. Patients having a single coronary artery, which arises from the aorta, have also been described. This anomaly, in contrast to anomalies of origin of these vessels, is compatible with a completely normal life span. From this evidence and anatomic studies Edwards and Burchell and Brown postulated that anomalous origin of a coronary artery produced myocardial ischemia from inadequate blood supply to the heart and suggested that the proper operation would be ligation of the anomalous left coronary artery at its origin. This has led to the satisfactory treatment of many such infants in whom the collateral circulation between the two coronary arteries is adequate.

The clinical picture of this anomaly beyond infancy and in adults may vary from complete absence of symptoms to severe, incapacitating angina. Frequently, such patients are thought to have valvular heart disease because of the presence of a murmur from the fistula itself or from mitral regurgitation. Insufficiency of the mitral valve can be produced by this anomaly, with progressive cardiac dilatation and failure from ischemia, infarction, and fibrosis.

The question arises whether any patient with an anomalous left coronary artery who has survived the transitional period of infancy and developed adequate coronary collaterals should be treated surgically. The oldest patient previously treated by ligation of the anomalous artery was 16 years old, and she developed ischemia after operation (as judged by electrocardiographic changes). This patient had been asymptomatic before ligation. This introduced a note of caution in the management of these patients and suggested that ligation may not be a completely desirable procedure, in spite of obliteration of the arteriovenous shunt and the increase in pressure in the left coronary artery distal to the ligation. However, the small number of adults who have been found to have this anomaly, either at autopsy or by antemortem study, have frequently died suddenly from probable acute arrhythmias.

The present patient was 45 years old and had severe and incapacitating symptoms. Because of the progression of her difficulty and the extensive collateral vessels demonstrated by cineangiography, surgical treatment seemed necessary and reasonable. Two approaches were considered: one being simple ligation of the vessel at its origin, and the other, a vein autograft from the ascending aorta to the anomalous artery, with ligation of the artery at its origin. When occlusion of the fistula into the pulmonary circulation was found to raise the pressure in the anomalous vessel to that of the aorta, it was elected to perform ligation alone. With this elevation in pressure, it seemed unlikely that a bypass graft would further increase the blood supply to the left coronary system. No further ischemic changes were seen by electrocardiography postoperatively, and there were no enzyme changes suggesting myocardial necrosis. There was atrial arrhythmia postoperatively, which initially caused no difficulty. The episode of asystole clearly seemed to be due to cardiac depression from the inadvertent simultaneous and rapid intravenous infusion of potassium chloride and Xylocaine.

This case, therefore, not only indicates that older patients with this anomaly can be treated by obliteration of the fistula, but also suggests that there may be a fourth phase of the anomaly in addition to the three suggested by Edwards and Talner and associates. This stage could be called a "coronary steal syndrome." The arteriovenous fistula with run-off into the pulmonary artery which is present in the third phase could result in progressive dilatation of the left coronary system and its communications with the right side. The large pressure drop across the system would diminish blood
flow to the small coronary branches, and more blood could be shunted away from the myocardium and directed into the pulmonary circulation. This stage could be present shortly after adequate collaterals have developed in childhood but could also occur later in life, as seen in the present patient. In the third phase, as satisfactory collaterals develop, such a patient frequently becomes asymptomatic, but in a fourth phase symptoms of myocardial ischemia could again be present due to an increase in size of the arteriovenous fistula and stealing of blood from the left coronary distribution. The present patient had no symptoms until the age of 16, when she developed angina. This could have been due to degenerative changes in the coronary vessels at that time, but none were evident when she was studied 29 years later. The development of symptoms could also have been related to her growth. It seems more likely that the A-V shunt increased to produce symptoms, representing another stage of progression of this anomaly.

Studies of coronary artery distribution in normal individuals have shown different patterns of dominance of the right or left arteries. In 50% of hearts studied the right coronary artery was dominant so that this vessel supplies the apex or crux of the heart. In 25% the distribution is balanced so that both vessels supply the apex, and in the remaining 25% the left coronary artery is dominant.30 These variable patterns occurring in patients with anomalous left coronary arteries could influence whether or not adequate collaterals develop. Thus the pattern of coronary artery distribution in an individual with an anomalous artery may determine the various syndromes or "phases" which then are not altered. This potential influence must remain speculative until more detailed studies can be made.

When mitral regurgitation has been present along with an anomalous coronary artery which has not been diagnosed preoperatively, repair of the mitral valve alone has been disastrous. In patients with mitral regurgitation, the effects of ligation of the anomalous artery on heart size are not yet well enough established to determine whether ligation alone or ligation with mitral valve repair or replacement is required. In the present patient, the apical systolic murmur decreased gradually and disappeared after operation. This suggests that mitral regurgitation was present which then decreased.

References

Circulation, Volume XXXVI, December 1967
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Circulation. 1967;36:878-885
doi: 10.1161/01.CIR.36.6.878

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