The Direction of Blood Flow in Anomalous Left Coronary Artery Arising from the Pulmonary Artery

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MYOCARDIAL infarction in infancy secondary to the anomalous origin of the left coronary artery from the pulmonary artery is now a condition that is being recognized with increasing frequency during life. One of the more interesting aspects of this congenital abnormality is the problem of the direction of the blood flow in the left coronary artery. In 1886 St. John Brooks first postulated that the flow in an anomalous coronary artery was retrograde. Following an anatomic dissection in 2 cases in which the right coronary artery arose from the pulmonary artery, he concluded that arterial blood flowed from the normal left coronary artery, which arose from the aorta, through anastomotic collateral vessels into the right coronary artery with ultimate drainage into the pulmonary artery. If the flow is retrograde, it is apparent that one of the coronary arteries is not only failing to supply the myocardium but is actually draining fully oxygenated blood from the heart into the pulmonary artery. Recently Edwards and others have presented further evidence in support of this view. The present communication concerns a patient with anomalous origin of the left coronary artery from the pulmonary artery in whom the direction of blood flow in this vessel was studied and demonstrated conclusively.

Case Report

T. L. (B-65692). This 2½-month-old male infant was referred to the Harriet Lane Home Cardiac Clinic of the Johns Hopkins Hospital on June 3, 1959. He was originally examined at the Memorial Hospital of the University of North Carolina where a diagnosis of an anomalous origin of the left coronary artery from the pulmonary artery had been made by Drs. Herbert Harned and George Summer. The family history was not contributory. The child had a 4-year-old sibling, who was healthy and normal. The mother had noted considerable vomiting toward the end of the pregnancy and her weight gain had been 28 pounds. Delivery was performed by cesarean section 12 days prior to term and an excessive amount of amniotic fluid was noted. The birth weight was 7 pounds 11 ounces. He was a slow feeder from the beginning, and had slightly grunting respirations. An examination at the end of 1 month was said to have been normal. When the infant was approximately 6 weeks of age, he suddenly became quite red in the face and vomited during a feeding and was dyspneic for a short time. Approximately 2 weeks prior to admission he became irritable and shortly thereafter had a deep reddish-blue color of the skin. He was very dyspneic and vomited several times. Shortly thereafter the attending pediatrician noted a heart murmur. Chest films were interpreted as showing cardiomegaly, and the child was referred to the Memorial Hospital of the University of North Carolina. Marked cardiac enlargement was noted, and a grade-I systolic murmur was present along the left sternal border and at the apex. An electrocardiogram showed sinus tachycardia and the pattern of a recent anterior myocardial infarction. A cineangiogram showed the left ventricle to be markedly enlarged, with a very thin wall. There was no evidence of an intracardiac shunt. A diagnosis of anomalous origin of the left coronary artery from the pulmonary artery was made.

On admission to the Johns Hopkins Hospital the infant's weight was 11 pounds, 11 ounces. He appeared well developed and well nourished. He was active, alert, and without cyanosis. The respirations were 44 per minute and had a grunting quality, but there was no marked respiratory distress. The heart was enlarged to the left anterior axillary line and the sounds were somewhat distant. The second aortic and pulmonary sounds were equal. A grade-II, slightly harsh, systolic murmur was heard along the left sternal border and radiated over the precordium. The blood pressure in the right arm by the flush technic was
The femoral pulses were easily palpable. The lungs were clear to percussion and auscultation. The liver was palpable 1.5 cm. below the right costal margin and the spleen was felt 2 cm. below the left costal margin. The remainder of the examination was not remarkable.

Laboratory data: The hematocrit value was 32, the hemoglobin 10.1 Gm. per 100 ml. and the leukocyte count was 13,550 per mm. An electrocardiogram (fig. 1) showed sinus tachycardia at 140 per minute, left axis deviation, deep Q in leads I, aV_L, V_4, and V_5, elevation of the S-T segments and inverted T waves in V_5 and V_6. The diagnosis was made of recent anterior infarction.

A review of the chest films showed marked cardiomegaly with normal pulmonary vascular markings (fig. 2). The cineangiocardiogram revealed a small right heart with a massively dilated left ventricle. The left ventricular wall was quite thin and pulsations were barely discernible. No evidence of filling of the left coronary artery from the pulmonary artery was seen.

The diagnosis of anomalous origin of the left coronary artery from the pulmonary artery was confirmed and it was decided that operation should be performed. On June 6 the infant was lightly anesthetized with cyclopropane and a left anterior thoracotomy was performed with entrance into the chest through the fourth intercostal space. A considerable amount of fluid escaped from the pericardium. The left ventricle was greatly dilated. A definite myocardial infarct was present on the anterolateral surface of the left ventricle measuring approximately 3 by 4 cm. (fig. 3). With each beat of the heart the area of the infarct bulged paradoxically. A chest retractor was poorly tolerated, producing arrhythmias, bradycardia, and hypotension. It was removed and most of the operative procedure was done without it. The left coronary artery was dissected and was found to arise from the pulmonary artery. It was a vessel of normal size and its anterior descending and circumflex branches were also visualized. The pressure in the left coronary artery was recorded.

Figure 1

*Electrocardiogram at age of 2 months.*
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at 30/15 mm. Hg. An arterial clamp was then applied to the proximal portion of the artery at its origin and the distal pressure was found to be 75 mm. Hg systolic. A simultaneous mean pressure from the pulmonary artery was 25 mm. Hg. A sample of blood from the anomalous left coronary artery was found to have 100 per cent oxygen saturation. At the same time saturation of a sample from the pulmonary artery was 76 per cent. These observations were considered to demonstrate conclusively that the flow in the anomalous coronary artery was retrograde into the pulmonary artery. The anomalous artery was then ligated with 3 suture ligatures (fig. 3A) proximal to its division into anterior descending and circumflex branches. At this time the systolic systemic arterial pressure rose from 90 to 120 mm. Hg and it was thought that movement of the left ventricular infarct was less paradoxical. Concentrated phenol was then applied to the entire surface of the heart for de-epineuralization. The pericardium was left open and the chest was closed with catheter drainage of the pleural cavity. The patient tolerated the procedure well. An electrocardiogram was recorded continuously during the procedure and no significant or persistent changes occurred. The presumed course of blood flow in the left coronary artery before and after ligation is shown in figure 3B.

Following operation the child made an uneventful recovery. The mother noted a marked difference in his appetite and in ability to eat without respiratory distress. Six months after operation his physician reported that he is gaining weight and doing well. A recent electrocardiogram definitely showed less evidence of left ventricular myocardial ischemia, and a chest film showed a slight decrease in the size of the heart.

Discussion

In 1911 Abrikossoff3 first reported anomalous origin of the left coronary artery from the pulmonary artery. Little interest was shown in this condition except for occasional reports until 1933 when Bland, White, and Garland4 first recorded an electrocardiogram in an infant with this condition and described the clinical and pathologic features. More than 60 cases are now recorded in the literature.

Early diagnosis of anomalous origin of the left coronary artery from the pulmonary artery is essential if successful therapy is to be employed. In many infants with this anomaly myocardial damage may become so extensive that the changes become largely irreversible within a relatively short period. For this reason early recognition and diagnosis is imperative if the most successful results are to be obtained by surgical therapy. The condition should be suspected in an infant having cardiomegaly associated with electrocardiographic evidence of left ventricular infarction. Further confirmation may be obtained by angiocardiography with demonstration of a normal right ventricle and a massively enlarged, thin-walled, left ventricle. It has been noted previously that the anomalous left coronary does not usually fill from the pulmonary artery when contrast media are injected into the right side of the heart, but the normal right coronary artery arising from the aorta may be demonstrated by the performance of a retrograde aortogram.

The seriousness of this anomaly is emphasized by the fact that in untreated cases death secondary to myocardial infarction and cardiac failure usually occurs within the first 5 months. An excellent review of the clinical manifestations and course of infants with this anomaly has been published recently by Keith.5

In the past various forms of surgical therapy have been advocated. Gasul and Loeffler6

Figure 2
Preoperative chest film demonstrating marked enlargement of heart, particularly of the left ventricle.
first suggested the creation of an aortic-pulmonary fistula in an effort to increase the arterial pressure and oxygen content of the blood in the pulmonary artery. This was attempted by Potts in 1921 on two occasions but was unsuccessful. In 1955 Kittle and associates planned a constriction of the pulmonary artery just distal to the origin of the anomalous left coronary artery in an effort to increase the pressure within it, but the infant died while the incision was being made. Others have recognized that the ideal surgical approach to this condition is transplantation of the anomalous coronary artery into a systemic artery by direct anastomosis. Such a procedure is necessarily tedious and difficult, and infants with this condition have an extremely limited myocardial reserve. This procedure was attempted by Mustard in 1953 but the child did not survive. Apley and associates advocated resection of the infarct as a means of reducing the paradoxical motions of the left ventricle as well as ligation of the left coronary artery. Paul and Robbins have employed talc poudrage and recommend its use. In our own experience 2 infants have been treated by chemical de-epicardialization (phenol) in an effort to revascularize the left ventricle. This procedure has appeared to provide real benefit, since both children are surprisingly well 2 and 4 years following operation although cardiomegaly persists.*

Within the past several years there has been increased interest in the direction of blood flow in an anomalous coronary artery arising from the pulmonary artery. In 1886 St. John Brooks reported observations on 2 hearts with this condition, which he found incidentally in the anatomic dissecting laboratory. In both instances the right coronary artery arose from the pulmonary artery and his dissections led him to make the following observation: "A consideration of this case will show that a very interesting question is connected with it. Here are two arteries belonging to the different circulations—the pulmonary and the systemic anastomosing with each other. In these circumstances, as is well known, the arterial pressure is very much greater in the systemic than in the pulmonary; how then did the blood flow in the anomalous coronary artery? There cannot be doubt that it acted very much after the manner of a vein, and that blood flowed through it towards the pulmonary artery, and from thence into the lungs." Later, Maude Abbott also endorsed this theory of retrograde flow of blood under these circumstances. Recently Edwards has renewed interest in a reconsideration of this problem and has called attention to the fact that infants with congenital cyanotic cardiac disease often have an arterial oxygen saturation which is below that of the venous blood in patients without right-to-left shunts. Yet these patients do not show clinical or pathologic evidence of myocardial ischemia. For this reason he concludes that the reduced perfusion pressure in the anomalous left coronary artery is the most important feature, and has advocated ligation of this vessel as the therapeutic method of choice. This has been performed by Morrow, Jahne, and others. Edwards offered the following points in support of the theory of retrograde flow: (1) In both normal and anomalous coronary arteries anastomoses exist between the branches; (2) in anomalous origin of the left coronary artery from the pulmonary artery both the right and left coronary arteries have been observed to have been extremely dilated and tortuous in a manner

*These infants will be reported separately as a part of our total experience with this condition.

Figure 3

A. Illustration of heart at operation showing area of infarction and aneurysmal dilation of left ventricle. The left coronary artery was ligated at its origin. B. Direction of blood flow in anomalous coronary artery. Before ligation oxygenated blood from the normal right coronary artery under systemic arterial pressure flowed through collateral vessels into the left coronary with retrograde drainage into the pulmonary artery. After ligation the arterial blood was shunted to the peripheral circulation of the left coronary artery where it could be used by the myocardium.
similar to that occurring in other situations in which arterial venous communications are known to be present; (3) clinical evidence of myocardial ischemia is usually not apparent until several months after birth at a time when the difference between the pressures in the systemic and pulmonary circuits become established; (4) perfusion studies on post-mortem specimens with these anomalous vessels show evidence of large communications between the anomalous and normal coronary arteries; and (5) an observation by Apley and associates that when the coronary artery which arose from the pulmonary trunk was divided at operation bright red blood flowed freely from its distal end."

We have also been able to demonstrate free communication between the left and right coronary arteries in 2 autopsy specimens of the heart from infants with anomalous origin of the left coronary artery from the pulmonary artery. Radiopaque contrast media injected into the right coronary artery completely filled the left coronary artery immediately and actually prior to the filling of the coronary veins. Similarly, methylene blue injected into the right coronary artery quickly filled the left coronary artery and was noted to flow into the pulmonary artery in significant amounts.

In the present case it was possible to isolate the anomalous coronary artery that arose from the pulmonary artery and to measure the pressure within it. This was first done with the vessel open and later with it occluded at its origin. With proximal occlusion of the vessel the pressure rose sharply in the artery, thus suggesting strongly that the source of blood was from the collateral branches. In addition, a sample of blood drawn from the anomalous artery showed full oxygen saturation (100 per cent), whereas a simultaneous sample from the pulmonary artery showed a saturation of 76 per cent. These observations appear to demonstrate conclusively that the blood in an anomalous left coronary artery arising from the pulmonary artery does in fact flow in a retrograde manner. It lends further support to the concept that ligation of the vessel at its origin is a rational method of treatment. This procedure was performed in the infant discussed in the present communication with apparent success.

**Summary**

An infant with anomalous origin of the left coronary artery from the pulmonary artery is presented, and the problem of the direction of blood flow in the anomalous vessel is discussed. Determinations of arterial pressure and oxygen saturation in the blood of the anomalous artery have demonstrated that the blood in this vessel flowed in a retrograde manner. It is concluded that in this condition oxygenated blood flows from the aorta into the normal right coronary artery and passes through collateral branches into the left coronary artery with ultimate drainage of the blood into the pulmonary artery. Ligation of the anomalous coronary artery with concomitant de-epicardialization appears to be a rational and effective method of therapy. For maximal effectiveness the operation should be performed prior to the onset of irreversible changes, and emphasis is placed upon the importance of early diagnosis and prompt treatment.

**Summario in Interlingua**

Es presentate le caso de un infante con un arteria sinistro-coronaria a origine anormal in le arteria pulmonar. Le problema del direction del fluxo in le vaso anormal es discutite. Determinaciones del tension arterial e del saturation de oxygeneo in le sanguine del anormal arteria ha servite a demonstrar que le fluxo del sanguine in iste vaso sequene un direction retrograde. Es concludite que in iste condition sanguine oxygenate flu in le aorta in le normal arteria dexter-coronaria e passa per braneas collateral in le arteria sinistro-coronaria, resultante ultimemente in un drainage del sanguine in le arteria pulmonar. Ligation del arteria coronaria anormal in concomitantia con dis-epicardialisation pare esse un plausibile e efficace metodo therapeutic. Pro esser maximalmente efficace, le operation deberea esser effectuate ante le declaration de irreversible alterationes. Es subliniate le importantia de un precoce diagnosto e de un prompte tratamento.

**References**

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It cannot be a subject of surprise that, with the above opinions, acquired chiefly during the latter period of his life, he should have retracted, in his second edition, the much more accurate doctrines respecting bellows-murmur as a sign of valvular disease, which he had advanced in his first;—transmitting to his disciples the confusion which reigned in his own mind, but which, like the storm that, in tropic climes, is the precursor of the purest, brightest weather, must, sooner or later, had his life been spared, have rolled away before the irresistible force of his purifying and enlightening genius.—J. Hope, M.D. Diseases of the Heart and Great Vessels. London, William Kidd, 1832, p. 16.
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