Anomalous Left Coronary Artery Originating from the Pulmonary Artery

Report of Two Surgically Treated Cases with a Proposal of Hemodynamic and Therapeutic Classification

By Alexander S. Nadas, M.D., Raúl Gamboa, M.D.,
and Paul G. Hugenholtz, M.D.

Recent reports have indicated that anomalous coronary arteries originating from the pulmonary artery may act as a channel for left-to-right shunt.¹-⁴ Thus a large portion of the coronary blood flow may bypass the coronary capillary bed, pass through arterial anastomoses, and enter directly into the pulmonary artery by retrograde flow through the anomalous coronary with resultant myocardial ischemia. In infants suspected of having this anomaly, early death has been ascribed to this mechanism. This has resulted in a strong plea for early ligation of the aberrant coronary artery in an attempt to redistribute the coronary flow throughout the entire capillary bed.¹ The observation of successful ligation of the aberrant vessel in one case, with prolonged postoperative follow-up, and of sudden death after the same procedure in another, has led to a revised evaluation of this problem and has prompted this report.

Case Reports

Case 1

A.J., a 5-week-old boy, was the product of a full-term uncomplicated pregnancy and normal delivery. There was no family history of heart disease. The patient’s birth weight was 6 pounds 9 ounces and he apparently did well until 12 hours prior to admission when he began to vomit and became tachypneic and febrile.

On admission the patient was acutely ill, with rapid, shallow, grunting respirations. The heart rate was 160 and regular. Peripheral pulses were of good quality. The first sound was increased in intensity, the second sound was louder than normal. There was a grade-II blowing systolic murmur at the apex, transmitted to the axilla. The liver was 3 cm. below the right costal margin. The lungs were clear, and there was no edema.

Urinalysis and hemogram were within normal limits. Chest films showed irregular areas of over-aeration of the lungs as well as marked accentuation of the pulmonary markings. There was considerable cardiac enlargement of all chambers, although the left side was predominant. A film 4 days later showed marked atelectasis of the right upper lobe, which persisted in subsequent films (fig. 1). The electrocardiogram showed an axis of -20° in the frontal plane projection, abnormal Q waves in leads I, aV₂, V₃, V₅, V₆, and a decreased magnitude of the R wave from V₄ to V₄R to V₂. The T waves were negative in leads I, aV₂, V₅, and V₆ (fig. 2).

On the basis of these findings, the diagnosis of pneumonia and anterolateral myocardial infarction with cardiomegaly due to an aberrant left coronary artery was strongly suspected. After recovery from the pneumonia, ligation of the aberrant coronary was recommended.

At thoracotomy an anomalous left coronary artery, originating from the posterior portion of the pulmonary artery, was found. The myocardium of the lateral portion of the left ventricle appeared cyanotic and edematous. The anomalous left coronary artery was ligated near its origin from the pulmonary artery. During the course of the operation, prior to the application of the ligature, ventricular fibrillation occurred and shortly after the anomalous vessel had been ligated, ventricular standstill took place. Resuscitation was successful on both occasions.

Following operation the child made an uneventful recovery with rapid weight gain. A
recent electrocardiogram taken 3 years after operation, showed persistence of the left axis deviation but disappearance of signs of myocardial infarction (fig. 2). The child is presently completely asymptomatic and is fully active.

Case 2
M.R.R., a 6-month-old girl, was the product

Figure 1

Case 1. Serial roentgenograms. Operation was performed after A and before B; C was obtained 2 years after surgery.
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Figure 2

Case 1. A, preoperative electrocardiogram. B, tracing of the same patient 2 years postoperatively. (Note the diminution of the Q waves in leads I, aVL, V1, V2, and V6 and persistence of -30° frontal plane axis).

of a full-term uncomplicated pregnancy and normal delivery. There was no family history of heart disease. The patient appeared well at birth, weighing 6 pounds 1 ounce. At 3 months of age she had an upper respiratory infection after which she failed to gain weight, sweated profusely with feedings, and showed intermittent cyanosis. The family physician noted poor color, a fast heart rate, and cardiomegaly without evidence of congestive heart failure. She was hospitalized and treated with digitalis. After some improvement and weight gain, digitalis was discontinued, against advice, and recurrent symptoms led to readmission.

The patient was irritable, with a high-pitched cry and dusky appearance. She weighed 11 pounds 11 ounces. The respiratory rate was 60. The heart rate was 150 and regular. Brachial and femoral pulses were of good quality. The first heart sound was normal at the apex, the second heart sound was closely split and normal in intensity, and a faint third sound was heard at the apex. No murmurs were present. The liver was 4 cm. below the right costal margin. The lungs were clear and there was no edema.

Urinalysis and hemogram were within normal limits. Fluoroscopic and radiographic examination showed predominantly left ventricular enlargement (fig. 3). The cardiac beat showed marked diminution in amplitude along all borders.

Figure 3

Case 2. Posteroanterior and right oblique roentgenograms demonstrating marked enlargement of all chambers of the heart.
The main pulmonary artery and the pulmonary vasculature were not remarkable; the aortic arch was on the left.

The electrocardiogram showed an axis of $-50^\circ$ in the frontal plane projection, abnormal Q waves in leads I, aV_L, V_5, V_6, and a decreased magnitude of the R wave from V_2 to V_4. The T waves were negative in leads I, aV_L, V_5, and V_6. The vectorcardiogram by the Frank corrected lead system showed an abnormal clockwise rotation of the horizontal and frontal plane projections. In the horizontal plane the direction of the 0.01-, 0.02-, 0.03-, and 0.04-second vectors was 114°, 90°, 223°, and 266°, respectively. The T loop presented an anterior displacement (fig. 4).

These findings indicated the presence of severe anterolateral myocardial infarction with cardiomegaly and no definite evidence for congestive heart failure. Thus in this 6-month-old child an aberrant left coronary artery was suspected.

Cardiac catheterization showed normal pressures in the right side of the heart and no evidence of a left-to-right shunt by oxygen saturation data, hydrogen curves, and selective cineangiography with injection above the aortic valves (table 1 and fig. 5). The cineangiograms did show, however, an unusually large right coronary artery supplying the right side of the heart. No collateral vessels were seen, nor was a left coronary artery visualized in several injections. Contrast material delivered into the main pulmonary artery failed to show any coronary artery. These findings were compatible with the clinical diagnosis, but did not give any evidence as to the site of origin of this vessel and did not indicate the presence of a left-to-right shunt.

In view of the almost uniformly poor prognosis of patients with abnormal left coronary artery and the good result in case 1 and similar reports in the literature, a thoracotomy was undertaken to ligate the left coronary artery at its origin from the pulmonary artery, in spite of lack of proof for a left-to-right shunt. At operation an aberrant coronary artery originating from the pulmonary artery was found. The color of this vessel was blue. It was ligated and immediately severe ST-
Table 1

<table>
<thead>
<tr>
<th>Catheterization Data</th>
<th>Pressures (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen saturation (%)</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>a = 13, m = 6</td>
</tr>
<tr>
<td>Right atrium</td>
<td>a = 12, m = 4</td>
</tr>
<tr>
<td>Right ventricle, outflow</td>
<td>38/8</td>
</tr>
<tr>
<td>Right ventricle, low</td>
<td>a = 12, m = 9</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>27/12, m = 14</td>
</tr>
<tr>
<td>Aorta</td>
<td>83.4</td>
</tr>
<tr>
<td>Pulmonary capillary</td>
<td></td>
</tr>
<tr>
<td></td>
<td>a = 12, m = 9</td>
</tr>
</tbody>
</table>

Assumed oxygen consumption: 200 ml./min./M.²
Systemic flow: 7.4 L./min.
2.1 L./min./patient
Hydrogen arrival time: pulmonary capillary = 1 second
Pulmonary artery and right ventricle; no early arrival on multiple curves

segment elevation was seen on the electrocardiogram (fig. 6). Ventricular standstill occurred twice, once during induction and once after removal of the endotracheal tube, but the heart beat was easily restored. At this time the patient seemed stable and was transferred to hyperbaric oxygen environment. Gradual decompression was begun and while the patient seemed to be doing well, sudden cardiac arrest occurred again. This time resuscitative measures were of no avail.

Autopsy revealed the ligature around an anomalous left coronary artery arising from the pulmonary artery, with recent myocardial infarction involving the lower half of the anterior wall of the left ventricle. An old myocardial infarction involving the anterolateral wall of the left ventricle was present as well (fig. 7).

Discussion

Anomalous origin of the left coronary artery, arising from the pulmonary artery, has become more frequently recognized clinically in recent years. Generally, two types have been distinguished in the approximately 75 cases reported in the literature. The rare adult form, in which the anomalous left coronary artery acts as a site of left-to-right shunting via multiple collateral vessels in the myocardium, is unassociated with electrocardiographic evidences of myocardial infarction and has a reasonable prognosis. On the other hand, in the common infantile form, in which such collateral flow is very poor or nonexistent, early death via severe myocardial ischemia is the rule.

The difference in the clinical course of these two types is almost certainly based on the number of collateral vessels remaining available for perfusion of the left ventricle. The distinction of adult versus infantile, therefore, reflects solely the degree to which the coronary collateral vessels have been effective in this regard.

Although in patients with the “adult type” anomaly, without myocardial infarction, the presence of a coronary artery-to-pulmonary artery shunt has been demonstrated repeatedly, there is no agreement about the hemodynamic status of infants with anomalous left coronary artery showing the electrocardiographic pattern of myocardial infarction. In our first patient, the clinical signs and the electrocardiographic studies strongly indicated the presence of an aberrant left coronary artery arising from the pulmonary artery with

Figure 5

Coronary angiograms after injection of contrast medium in ascending aorta. Patient in sharp left anterior oblique position. Only the right coronary artery fills and is of large caliber.
previous myocardial infarction of the left ventricular wall. The excellent improvement after ligation of this aberrant vessel indicated that the bypass of oxygenated blood from the normal right coronary artery into the pulmonary artery was stopped and that a reasonable left ventricular myocardial perfusion from the right coronary artery through collateral vessels was established.

In the second infant, the clinical, electrocardiographic, and vectorcardiographic studies indicating anterolateral myocardial infarction were also strongly suggestive of the presence of an aberrant left coronary artery from the pulmonary artery. Attempts accurately to delineate the origin and course of the aberrant vessel as well as the direction of blood flow in it, by means of oxygen saturation, hydrogen curves, and selective angiocardiology with injection into the ascending aorta and pulmonary artery, while not diagnostic, indicated no left-to-right shunting. Nevertheless, because of the widely accepted theory that the basic problem in these infants is that of bypassing of oxygenated blood from the normal right coronary artery into the pulmonary artery via the left coronary, as well as the favorable experience with the first case, we recommended ligation of the aberrant coronary artery, presuming that catheterization data had not been sensitive enough to demonstrate the left-to-right shunt. The course of this patient during and after surgery suggests that this was not a problem of left-to-right coronary shunting but rather that, because of the absence of collateral vessels between right and left coronary arteries, the left coronary artery alone supplied a large portion of the left ventricular myocardium, albeit with poorly oxygenated blood from the pulmonary artery at a low perfusion pressure. The frequency with which the left coronary artery is the sole supplier of the left ventricular myocardium is unknown. It is known, however, that in 22 per cent of neonates examined post mortem with coronary injection studies no demonstrable collateral vessels between left and right coronary arteries were present. Furthermore, there is a case similar to ours that demonstrates a rapidly deteriorating course of the patient when a ligature was placed around the aberrant left coronary artery. Case and co-workers, as well as Lampe and Verheugt, have suggested that the effects of temporary occlusion of the aberrant left coronary artery should be evaluated at the time of operation prior to permanent ligation.

It would seem, then, that patients with anomalous origin of the left coronary artery
represent, like so many other congenital cardiac deformities, a spectrum. At one extreme are the patients with the “adult form” of the disease without clear-cut evidences of myocardial infarction, in whom the right coronary artery, through an ample collateral network, drains into the left coronary artery and from there into the low-pressure pulmonary artery. This hemodynamic situation may well be demonstrated most clearly by means of selective arteriography. In effect, these anomalies may be regarded as representing coronary arteriovenous fistulas. Surgery, consisting of ligation of the left coronary artery at its entry into the pulmonary artery is recommended, to avoid low-grade myocardial damage, the flow work inherent in any left-to-right shunt, infection, and even possible rupture of the aneurysm.

At the other end of the spectrum are the infants without appreciable collateral network between right and left coronary artery, with the most critical disease, in whom the sole blood supply to the left ventricular myocardium is from the pulmonary artery through the anomalous left coronary artery. The low-pressure perfusion of the left ventricle with poorly oxygenated blood results in myocardial

Figure 7
Case 2. Photograph of postmortem specimen. A, the single coronary ostium arising from the aorta indicated by the arrow. Myocardial infarction is present in the left ventricular wall. B, anomalous left coronary artery arising from the main pulmonary artery indicated by the arrow.
infarction. Ligation of the anomalous left coronary artery in these critically ill infants deprives the left ventricle of its only blood supply and results in instantaneous death from myocardial ischemia. These infants with anterolateral myocardial infarction may probably be identified by the absence of any evidence of a left-to-right shunt from coronary artery to pulmonary artery by the most sensitive methods available (oxygen saturation, hydrogen, radiopaque dye) and at the operating table by the low oxygen content of the left coronary artery blood as well as by exaggeration of the ischemic pattern in the electrocardiogram when the entrance of the coronary vessel into the pulmonary artery is partially occluded. Coronary artery ligation is catastrophic in these patients.

In between these two extremes of the spectrum, are infants with anomalous left coronary artery and myocardial infarction in whom a collateral network of some magnitude between right and left coronary artery is available, but the preferential flow of blood into the low-pressure pulmonary artery deprives the left ventricular myocardium of an adequate oxygen supply. These are patients who benefit maximally from left coronary artery ligation; this maneuver forces oxygenated right coronary artery blood into the left ventricular capillary network. These patients may be identified as suitable candidates for coronary artery ligation preoperatively by demonstration of a left-to-right shunt at the pulmonary artery level and by selective arteriography. At the operating table it may be seen that the left coronary artery carries red, well-oxygenated blood and that temporary occlusion of its orifice is tolerated without deterioration in the electrocardiogram.

Thus it would appear that preoperative catheterization studies are essential in every case of abnormal origin of the left coronary artery if the patient's condition permits. If a left-to-right shunt is demonstrated (from aorta to pulmonary artery through the coronary system), the operation of choice would be ligation of the left coronary near its origin. If no left-to-right shunt is demonstrable, the surgeon should be prepared to evaluate coronary flow at the time of operation. A cyanotic-appearing left coronary artery, which shows low oxygen saturation, suggests poor anastomoses between the right and left coronary artery system. If a temporary ligature placed around the aberrant coronary does not cause the proximal vessel to distend and if the myocardium does not appear better oxygenated and, particularly, if the electrocardiogram shows fresh evidence of acute myocardial ischemia, the ligature should be removed. There are a number of actual or theoretical alternatives at this point.

The patient might be benefited by procedures that would improve oxygenation of the myocardium either by supplying poorly oxygenated blood under higher pressure (i.e., banding of distal pulmonary artery) or by supplying more highly oxygenated blood at low pressure (i.e., Blalock procedure).

**Summary**

Case histories of two infants with anomalous origin of the left coronary artery from the pulmonary artery are presented.

Both infants underwent ligation of the left coronary artery at its entrance into the pulmonary artery. One survived the operation and is asymptomatic 3 years postoperatively, whereas the second one died at surgery.

A classification of instances of anomalous left coronary artery according to adequacy of the collateral circulation between the left and right coronary systems is proposed.

Surgical ligation of the pulmonary artery orifice of the left coronary artery is indicated only if the presence of a left-to-right shunt into the pulmonary artery is demonstrated. If the direction of blood flow is from pulmonary artery to left coronary artery, surgical procedures should be aimed toward increasing either pulmonary artery oxygen content or left coronary artery perfusion pressure.

**Acknowledgment**

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References


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William Withering

Withering's first use of foxglove in his practice was in 1775, the year before he left Stafford for Birmingham. How it was first called to his attention is of such interest that it is given here in his own words:

"In the year 1775 my opinion was asked concerning a family recipe for the cure of dropsy. I was told that it had long been kept a secret by an old woman in Shropshire who had sometimes made cures after the more regular practitioners had failed. I was informed also that the effects produced were violent vomiting and purging; for the diuretic effects seemed to have been overlooked. This medicine was composed of twenty or more different herbs; but it was not very difficult for one conversant in these subjects to perceive that the active herb could be no other than foxglove."

The knowledge of the use of foxglove in dropsy may have been known in household medicine in that part of England just as a knowledge that cowpox protected against smallpox was known in Gloucestershire long before the time of Jenner. It needed a man like Withering to study in a scientific manner the effects of this rural remedy and introduce it to the pharmacopeia just as Jenner had taken the countryside tradition of cowpox and by study and observation developed the practice of smallpox vaccination from it.—Louis H. Roddis, M.D. William Withering: The Introduction of Digitalis Into Medical Practice. New York, Paul B. Hoeber, Inc., 1936, p. 50-51.
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ALEXANDER S. NADAS, RAUL GAMBOA and PAUL G. HUGENHOLTZ

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