Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

A New Method of Surgical Repair

By William H. Neches, M.D., Robert A. Mathews, M.D., Sang C. Park, M.D., Cora C. Lenox, M.D., James R. Zuberbuhler, M.D., Ralph D. Siewers, M.D., and Henry T. Bahnson, M.D.

SUMMARY
Anomalous origin of the left coronary artery from the pulmonary artery has been surgically corrected by means of a technique that avoids direct suturing of the coronary artery itself. The anomalous left coronary artery is removed from the pulmonary artery with a cuff of pulmonary artery tissue about its orifice. In a five- and-one-half-year-old patient the anomalous coronary artery was anastomosed directly to the aorta using this technique. In another patient, seven months of age, the anomalous coronary artery could not be anastomosed directly to the aorta and a segment of the patient’s subclavian artery was interposed between the left coronary artery and the aorta to complete the anastomosis. In the latter patient, profound hypothermia and circulatory arrest were used during construction of the anastomosis in an attempt to minimize further myocardial damage. The establishment of a two coronary artery system has been demonstrated to be feasible in the infant or young child with this anomaly.

Additional Indexing Words:
Profound hypothermia
Coronary artery anomalies
Myocardial infarction

ANOMALOUS ORIGIN of the left coronary artery from the pulmonary artery usually presents during infancy or early childhood.1-3 The earliest surgical treatment consisted of ligation of the anomalous left coronary artery (ALC) which eliminated the left to right shunt and produced a one-coronary system.1-5 Establishment of prograde left coronary blood flow has become possible recently with aorto-coronary artery saphenous vein bypass grafts.4-9 Although this method has been utilized in older children and adults, its use in infants and small children is limited because of the small size of their coronary arteries. A technique for direct anastomosis of an anomalous right coronary artery to the aorta in an adolescent has recently been described.10 This report will describe modifications of the technique of Tingelstad et al.10 which have been employed at the University of Pittsburgh School of Medicine for correction of anomalous origin of the left coronary artery from the pulmonary artery in two children, one of whom was less than a year of age.

Case Reports
Case 1
A 5%-year-old Caucasian boy was referred to Children’s Hospital of Pittsburgh for evaluation of a heart murmur first heard during a routine physical examination six months previously. The patient’s growth and development had been normal and there were no symptoms of cardiac disease. On physical examination there was no cyanosis. The heart sounds were normal. A grade 2/6 harsh systolic ejection murmur was heard best at the mid left sternal border and was transmitted to the mitral area. An intermittent faint continuous murmur was present over the same areas. No other murmurs, clicks or gallops were heard. Peripheral pulses were equal and symmetrical.

Chest roentgenogram demonstrated mild cardiomegaly and slightly increased pulmonary vascular markings (fig. 1). The electrocardiogram and vectorcardiogram showed left axis deviation and ST segment and T wave changes (fig. 2).

Cardiac catheterization revealed no intracardiac shunting or hemodynamic abnormalities (table 1). Selective left ventricular cineangiography demonstrated a normally contracting left ventricle and a dilated tortuous right coronary artery which arose from the aorta in a normal position. The left coronary artery filled in a retrograde manner from the right through multiple collateral channels and then emptied into
the pulmonary artery (fig. 3). The cardiac catheterization diagnosis was anomalous origin of the left coronary artery from the pulmonary artery with adequate collateral circulation.

The patient underwent surgical correction of the anomaly in August 1973, one month following cardiac catheterization. The operation was performed through a median sternotomy using cardiopulmonary bypass and mild hypothermia (rectal temperature 30° C). The right coronary artery was large and tortuous and arose normally from the aorta. The left coronary artery originated from the postero-lateral wall of the main pulmonary artery above the left pulmonary sinus of Valsalva and divided into anterior descending and circumflex branches. After cardiopulmonary bypass was begun, the pulmonary artery was opened anteriorly and a large amount of blood flowed from the anomalous left coronary artery (ALC) which was then occluded externally. In an attempt to get optimal exposure, the pulmonary artery was transected just distal to the pulmonary valve. A small
cuff of pulmonary artery surrounding the orifice of the ALC was excised and could be easily approximated to the aorta. A circular 6 mm opening was made with an aortic punch into the left lateral surface of the aorta just above the commissures. The end of the left coronary artery with its cuff of pulmonary artery was then anastomosed to the opening in the aorta and the pulmonary artery reanastomosed. Cardiopulmonary bypass was discontinued without difficulty.

**Table 1**

<table>
<thead>
<tr>
<th>Catheter position</th>
<th>Case 1</th>
<th></th>
<th>Case 2</th>
<th></th>
<th>Case 3</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preop: Age 5-6/12</td>
<td>Postop: Age 5-10/12</td>
<td>Preop: Age 6/12</td>
<td>Postop: Age 8/12</td>
<td>Preop: Age 11/12</td>
</tr>
<tr>
<td>SVC</td>
<td>—</td>
<td>82 — —</td>
<td>—</td>
<td>63 — —</td>
<td>—</td>
</tr>
<tr>
<td>RA</td>
<td>m = 1</td>
<td>80 m = 5</td>
<td>75 m = 3</td>
<td>80 m = 5</td>
<td>75 m = 3</td>
</tr>
<tr>
<td>RV</td>
<td>24/0 ED = 3</td>
<td>77 25/0 ED = 4</td>
<td>76 45/0 ED = 7</td>
<td>64 54/0 ED = 7</td>
<td>64 54/0 ED = 7</td>
</tr>
<tr>
<td>PA</td>
<td>24/10 m = 12</td>
<td>95 25/10 m = 12</td>
<td>77 40/20 m = 30</td>
<td>64 40/20 m = 30</td>
<td>64 40/20 m = 30</td>
</tr>
<tr>
<td>PAW</td>
<td>m = 6</td>
<td>— m = 5</td>
<td>— m = 15</td>
<td>— m = 15</td>
<td>— m = 15</td>
</tr>
<tr>
<td>LV</td>
<td>110/0 ED = 5</td>
<td>94 110/0 ED = 5</td>
<td>95 85/0 ED = 20</td>
<td>95 85/0 ED = 20</td>
<td>95 85/0 ED = 20</td>
</tr>
<tr>
<td>AO</td>
<td>110/70 m = 90</td>
<td>94 110/70 m = 90</td>
<td>95 85/0 ED = 20</td>
<td>95 85/0 ED = 20</td>
<td>95 85/0 ED = 20</td>
</tr>
<tr>
<td>CI = 5.04 L/m²/m²</td>
<td>CI = 4.8 L/m²/m²</td>
<td>CI = 4.8 L/m²/m²</td>
<td>CI = 4.8 L/m²/m²</td>
<td>CI = 4.8 L/m²/m²</td>
<td>CI = 4.8 L/m²/m²</td>
</tr>
</tbody>
</table>

Abbreviations: AO = aorta; CI = cardiac index; ED = end-diastolic pressure; LV = left ventricle; LV dP/dt = rate of rise of left ventricular pressure; m = mean; PA = pulmonary artery; PAW = pulmonary artery wedge; RA = right atrium; RV = right ventricle; SVC = superior vena cava.
and the remainder of the operation proceeded uneventfully. The patient’s postoperative course was uncomplicated and he has remained free of symptoms during the subsequent 9 months. The ST segment and T wave abnormalities seen on the preoperative electrocardiogram and vectorcardiogram were no longer present. Cardiac catheterization was performed three months postoperatively (table 1). There was no left to right shunt and the hemodynamics were normal. Left ventriculography demonstrated normal findings. Selective coronary arteriography showed a normal sized right coronary artery and there was no evidence of collateral circulation. The left coronary artery was widely patent and filled from the aorta in a normal manner (fig. 4).

Case 2
This six-month-old black male infant was referred to Children’s Hospital of Pittsburgh because of congestive heart failure. His growth and development had been normal. At 5 months of age he was hospitalized at another in-
ANOMALOUS CORONARY ARTERY: NEW REPAIR

Figure 6

Case 2. Electrocardiogram and Vectorcardiogram. In the ECG, left axis deviation (axis - 30°) is present. Abnormal Q waves and T wave inversion in leads I, aVL, and V4 and loss of mid precordial anterior forces suggest an anterior myocardial infarction. In the VCG, the frontal plane loop is superiorly oriented and is clockwise. The horizontal plane loop is inscribed clockwise and is completely posterior. The lack of anterior, inferior and leftward forces indicates an anterior myocardial infarction.

Stithution because of “bronchitis.” Three weeks later he again became ill with tachypnea and respiratory distress. Chest roentgenogram showed cardiomegaly and the patient was referred for evaluation. On physical examination he was a normally developed and nourished black infant weighing 7 kg (60th percentile). Tachypnea and tachycardia were noted but the lungs were clear and there was no hepatomegaly. The apex beat was in the sixth intercostal space at the anterior axillary line. The heart sounds were normal but a protodiastolic gallop was heard. There was a grade 2/6 blowing pansystolic murmur at the apex. No cyanosis, clubbing or edema was found. Peripheral arterial pulses were normal. Marked cardiomegaly was seen on the chest roentgenogram (fig. 5). The electrocardiogram showed deep Q waves in the lateral precordial leads and loss of anterior forces suggesting anterior wall myocardial infarction. This was confirmed by the vectorcardiogram (fig. 6).

Cardiac catheterization was performed at six months of age (table 1). There was no evidence of a left to right shunt by oximetry. Selective cineangiocardioigraphy revealed a dilated left ventricle with generalized poor contractility but no localized akinetic or dyskinetic areas were seen. On aortography a dilated right coronary artery was seen to arise normally from the aorta. The left coronary artery filled by collateral circulation from the right and then emptied into the pulmonary artery (fig. 7). Cardiac catheterization diagnosis was anomalous origin of the left coronary artery from the pulmonary artery with secondary anterior wall myocardial infarction and severe left ventricular dysfunction.

At age seven months, in January 1974, surgery was performed through a median sternotomy using cardiopulmonary bypass and profound hypothermia with temporary circulatory arrest. At operation the left coronary artery was found to arise from the left lateral side of the pulmonary artery. Direct anastomosis to the aorta was not possible so a 2 cm segment of subclavian artery was excised for use as a graft.

The proximal branches of the left coronary artery were dissected free and under profound hypothermia (rectal temperature 17°C) and using cardiopulmonary bypass, the ALC was occluded and detached from the pulmonary artery with a cuff of pulmonary artery tissue about its orifice. After

Figure 7

Case 2. Preoperative aortogram. A. Initially only a large right coronary artery is seen filling from the aorta (AO). B. During a later phase a small left coronary artery (arrow) fills by collateral circulation and empties into the pulmonary artery (PA).
the pulmonary artery had been closed, one end of the segment of subclavian artery was anastomosed to the rim of pulmonary artery tissue around the orifice of the ALC. A clamp was then placed on the ascending aorta, cardiopulmonary bypass was stopped and the patient was exsanguinated into the pump. The aorta was opened and a small button of tissue removed from the left lateral side just above the usual position of the left coronary artery. The subclavian artery graft was then passed posterior to the pulmonary artery and its proximal end was anastomosed to the aorta (fig. 8). The aorta-subclavian artery anastomosis was 4 mm in diameter and the subclavian artery-ALC anastomosis was 3 mm in diameter. Cardiopulmonary bypass was reintroduced and the reaming phase as well as the remainder of the operation proceeded without difficulty.

Postoperatively the patient’s recovery was delayed by respiratory difficulties secondary to phrenic nerve paralysis; however the cardiovascular status remained stable. The electrocardiogram and vectorcardiogram were unchanged from the preoperative studies. Cardiac catheterization was performed two weeks postoperatively (table 1). Retrograde aortography demonstrated patency of the anastomosis between the left coronary artery and aorta (fig. 9).

Discussion

In the asymptomatic child or young adult with anomalous origin of the left coronary artery from the pulmonary artery and normal left ventricular function, the major advantage of using an aorto-coronary artery bypass graft, as compared to ligation of the ALC, would seem to be in the lessened hazard of subsequent atherosclerotic coronary artery disease. On the other hand, in the infant or child with small intercoronary collaterals and consequent poor myocardial function, perfusion of the left coronary artery system may be inadequate after ligation of the ALC.13,14 Since the prograde flow from aorta to left coronary artery following a saphenous vein bypass graft should increase perfusion pressure in the left coronary artery, it is reasonable to assume that this might improve function in the ischemic but noninfarcted regions of the left ventricle. Although this latter technique has been used successfully in older children and adults, its usefulness in infants and young children is limited because of the small size of their coronary arteries. This report has described a modification of a technique for repair of an anomalous coronary artery9 that enables its use in the small child since it avoids any direct suturing on the coronary artery itself.

Figure 8
Case 2. Surgical technique. a. Excision of the anomalous left coronary artery along with a cuff of pulmonary artery tissue. b. Closure of the opening in the pulmonary artery. c. Anastomosis between the subclavian artery segment and the cuff of pulmonary artery tissue about the orifice of the left coronary artery. d. The subclavian artery segment is passed behind the pulmonary artery and its proximal end anastomosed to a punch hole in the side of the aorta.

Figure 9
Case 2. Postoperative aortograms. There is filling of both the right and left coronary arteries. The aorta-left coronary artery anastomosis is widely patent. A. Antero-posterior view; B. Right anterior oblique view; C. Left anterior oblique view.
In case 1, the ALC was found to arise from the left postero-lateral side of the pulmonary artery enabling direct approximation of the ALC to the aorta. The removal of a cuff of pulmonary artery around the orifice of the ALC permitted a larger anastomosis than would otherwise have been possible. At cardiac catheterization three months postoperatively, the anastomosis was widely patent normally. In case 2 the ALC arose more laterally from the pulmonary artery so the surgical technique was modified to include a subclavian artery graft.

Profound hypothermia with circulatory arrest has not previously been used in correction of this anomaly. In case 2 it was used to minimize or prevent any further myocardial damage from hypoxia secondary to the cross-clamping of the aorta that was necessary during repair.

The majority of patients with anomalous origin of the left coronary artery from the pulmonary artery will present during infancy, as did case 2, with severe left ventricular dysfunction or myocardial infarction. Since there has been a high incidence of saphenous vein bypass graft failure in small children in the past, it has been recommended that infants and small children be treated either without surgery until an older age or by operative ligation of the ALC. However progressive clinical deterioration and myocardial damage may occur in these symptomatic patients as a result of their anomaly. Thus, if maximum benefit is to be obtained by establishing prograde coronary blood flow, this procedure should be done as early as possible, preferably as soon as the diagnosis is made. In case 2, it was hoped that increasing perfusion pressure in the left coronary artery system would improve myocardial function. After repair there was a fall in the left ventricular end-diastolic pressure and an increase in the rate of rise of the left ventricular pressure (LV dP/dt). This suggests that left ventricular function was improved although it was still abnormal.

The technique of excising a cuff of pulmonary artery tissue with the ALC obviates the need for direct suturing of small coronary arteries and permits a wider anastomosis than would otherwise be possible. In our two cases it has been shown to be effective in establishing normal prograde coronary artery blood flow even in the infant or young child. It is hoped that the use of a wide anastomosis will insure continued long term patency. The long range prognosis is unknown in an infant or small child who has already sustained myocardial damage prior to operation. Since corrective procedures for this anomaly heretofore have not been available for this group, the long term evaluation of patients offered correction under these circumstances will be necessary to establish the validity of its use.

References

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery: A New Method of Surgical Repair
WILLIAM H. NECHES, ROBERT A. MATHEWS, SANG C. PARK, CORA C. LENOX, JAMES R. ZUBERBUHLER, RALPH D. SIEWERS and HENRY T. BAHNSON

Circulation. 1974;50:582-587
doi: 10.1161/01.CIR.50.3.582

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1974 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/50/3/582