Anomalous Left Coronary Artery from Pulmonary Artery

Report of 11 Cases; Review of Indications for and Results of Surgery

By Lowell W. Perry, M.D., and Lewis P. Scott, M.D.

SUMMARY
Anomalous origin of the left coronary artery from the pulmonary artery has been noted in 11 patients at Children's Hospital of the District of Columbia from 1946 to 1969. Four of our cases plus 40 in the literature have been reported with complete catheterization data. These 44 infants and children can be divided into three groups (A, B, and C) according to the magnitude of the shunt at pulmonary artery level.

The type of treatment in infants and children with anomalous left coronary artery and increased oxygen saturation in the pulmonary artery at the time of catheterization (group A) appears to be immaterial, as all have survived. Mortality rates in patients with left-to-right shunts not large enough to be detected by oxygen studies (group B) indicate no significant difference in those treated medically or surgically; hence, if possible, surgery should be delayed in this group until after 1 to 2 years of age when surgical mortality is negligible. In patients with filling of the left coronary artery from the pulmonary artery (group C), direct anastomosis of the anomalous left coronary artery to the aorta seems to be the only therapeutic recourse.

To date, there are insufficient data to indicate the efficacy of direct anastomosis of the anomalous left coronary artery to the aorta in the sick infant.

Additional Indexing Words:
Myocardial infarction Congenital heart disease Ligation of coronary artery
Bland-White-Garland syndrome Anastomosis of coronary artery to aorta

A NOMALOUS ORIGIN of the left coronary artery from the pulmonary artery is a serious congenital cardiac defect associated with high mortality rates in infancy. While no long-term studies are available to document morbidity and mortality experience of survivors of surgery in infancy, congestive heart failure, arrhythmias, and sudden and premature death have been documented as complications of the untreated anomalous coronary artery in the adult patient.1-4

At the present, question exists as to when infants and children with this anomaly should have surgery performed and also about the type of operation. The purpose of this paper is to review the experience with this lesion at Children's Hospital of the District of Columbia and to review the reported experience to determine the indications for and results of surgical intervention in infants and children with this lesion.

Methods

The pathology files and cardiac registry of the Children's Hospital of the District of Columbia were perused. Eleven cases encountered from 1946
to 1969 were found (table 1). In nine (82%) of these 11 cases death had occurred and postmortem examination was performed. Six of the hearts were available for gross examination at this time. The pathology reports or clinical records of all patients were reviewed.

**Results**

**Pathologic Studies**

The anomalous left coronary artery arose from the left posterior pulmonary sinus in all nine specimens. The right coronary ostium was normal in seven. In two cases, an accessory right coronary ostium, adjacent to and smaller than the true coronary ostium, was also present.

The left ventricle was dilated and thickened in all specimens. Varying degrees of endocardial fibroelastosis were present. Gross areas of infarction of the anterolateral wall, particularly of the inner third portion, were noted. In two of the nine cases a left ventricular aneurysm was present.

One or both papillary muscles of the left ventricle were thickened and involved in the endocardial fibroelastosis process. The chordae tendineae generally were shortened. The mitral valve rings did not appear dilated, but the mitral valves were often thinned and had rolled edges.

Microscopic examination revealed myocardial fibrosis, degeneration, and congestion. Intracardiac calcification was noted in three specimens, and mural thrombosis in two.

A bicuspid right atrioventricular valve was present in one specimen and a patent ductus arteriosus in another, but otherwise there were no associated cardiac anomalies.

Gross and microscopic evidence of congestive heart failure was present in all specimens.

**Clinical Features**

Six patients were Negro, four were Caucasian, and one was Oriental. Of the seven
Table 2
Onset of Symptoms and Age of Death in Cases of Anomalous Left Coronary Artery from Pulmonary Artery

<table>
<thead>
<tr>
<th>Age (mo)</th>
<th>No. of patients in age group at Onset of symptoms (patients)</th>
<th>Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>1 - 1.9</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>2 - 2.9</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>3 - 3.9</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>4 - 4.9</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>5 - 5.9</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>6 - 6.9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>7 - 7.9</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>14 - 14.9</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>

females none survived, and of the four males two survived. One of the cases had been reported previously.5

Symptoms began when patients were from 3 weeks to 4 months of age with the greatest number experiencing onset of symptoms during the second month of life (table 2). Symptoms generally fell into two groups; those of heart failure—poor appetite, failure to thrive, dyspnea, and cyanosis—and those of pulmonary infection—fever, irritability, tachypnea, and cough.

Physical findings fell into three categories: failure to thrive, heart failure, and pulmonary infection. Failure to thrive was noted frequently, primarily in terms of weight. Of the 11 patients, six (55%) were in the third percentile or less for weight, three (27%) were in the 10th percentile, and two (18%) were in the 25th percentile. None was above the 25th percentile for weight. Of eight patients measured for length, none was below the third percentile, and half were between the third and 10th percentiles.

Signs of heart failure included tachypnea, tachycardia, irritability, diaphoresis, and weak cry. Marked hepatomegaly was noted in 70%. Half of the patients had mitral regurgitation. Cyanosis was present in at least 40% of the patients when initially examined and might have reflected either heart failure or pulmonary infection.

Wheezees, rales, and rhonchi were the primary signs of pulmonary infection although these could also indicate left heart failure. Fever was noted in 20% of the patients on initial examination. Bronchopneumonia was frequently diagnosed on admission.

X-rays

X-rays were obtained in 10 and were available for review in six of our cases. The reports of the other four were reviewed. The chest x-rays uniformly revealed cardiac enlargement. The left chest was often opacified as a result of marked cardiac enlargement, atelectasis of portions of the left lung, or passive pulmonary congestion. Left atrial and left ventricular enlargement were noted on all x-rays examined.

Electrocardiographic Studies

Electrocardiograms were obtained in eight of the 11 patients. The electrocardiograms of all eight patients were abnormal. There was evidence of anterolateral myocardial infarction in seven (88%) and of a posterolateral infarction in one (12%). In one patient, the sole postoperative survivor, there was only loss of R-wave voltage across the left chest initially; Q waves developed following surgery. Serial electrocardiographic changes indicating extension of infarction were noted in two other patients.

Table 3 reveals that the Q waves were wide and deep. The most outstanding finding was the depth of the Q waves in aVL. Other pertinent ECG findings are listed in table 4. One patient had electrocardiographic evidence of supraventricular tachycardia when she was seen initially.

Table 3
Amplitude and Duration of Q Wave in Anomalous Left Coronary Artery from Pulmonary Artery

<table>
<thead>
<tr>
<th>Q wave</th>
<th>I</th>
<th>aVL</th>
<th>V1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration (sec)</td>
<td>0.01-0.03</td>
<td>0.01-0.035</td>
<td>0.02-0.035</td>
</tr>
<tr>
<td>Amplitude (mm)</td>
<td>Mean</td>
<td>2.8</td>
<td>6.3</td>
</tr>
<tr>
<td>Range</td>
<td>1-4</td>
<td>4-12</td>
<td>2-6</td>
</tr>
<tr>
<td>Q/R × 100 (%)</td>
<td>Mean</td>
<td>13</td>
<td>82</td>
</tr>
<tr>
<td>Range</td>
<td>5-29</td>
<td>18-250</td>
<td>8-32</td>
</tr>
</tbody>
</table>
Table 4
ECG Findings in Anomalous Left Coronary Artery from Pulmonary Artery

<table>
<thead>
<tr>
<th>Condition</th>
<th>Cases</th>
<th>% of total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upright T wave, right chest</td>
<td>7</td>
<td>88</td>
</tr>
<tr>
<td>Flat or inverted T wave, left chest</td>
<td>3</td>
<td>38</td>
</tr>
<tr>
<td>S-T segment elevated, left chest</td>
<td>5</td>
<td>63</td>
</tr>
<tr>
<td>S-T axis frontal plane, normal</td>
<td>5</td>
<td>63</td>
</tr>
<tr>
<td>Right, inferior</td>
<td>2</td>
<td>25</td>
</tr>
<tr>
<td>Left, superior</td>
<td>1</td>
<td>12</td>
</tr>
<tr>
<td>Loss of R-wave amplitude V2-V6</td>
<td>5†</td>
<td>72†</td>
</tr>
<tr>
<td>Left atrial enlargement</td>
<td>4</td>
<td>50</td>
</tr>
<tr>
<td>Left ventricular enlargement</td>
<td>6</td>
<td>75</td>
</tr>
<tr>
<td>Supraventricular tachycardia</td>
<td>1</td>
<td>12</td>
</tr>
<tr>
<td>Q waves not present initially</td>
<td>1</td>
<td>12</td>
</tr>
</tbody>
</table>

*Evaluated in eight cases unless otherwise stated. †Evaluated in only seven cases.

The electrocardiogram of patient 11 when first seen in 1955 is reproduced in figure 1 as typical of the group with electrocardiographic evidence of an anterolateral infarction. Left ventricular hypertrophy was also present. Repeat electrocardiogram in 1968, 13 years after the initial infarction, still reveals the presence of a Q wave in leads I, aVL, and V6 (fig. 2). The amplitude of the Q wave has decreased markedly in leads I and aVL, but the duration is greater than could be considered normal. The Q wave in V6 could not be considered abnormal in this tracing. The S-T and T-wave changes noted in infancy have resolved. There is still voltage evidence of left ventricular hypertrophy.

Enzyme Studies

Levels of serum glutamic oxalacetic transaminase (SGOT), serum glutamic-pyruvic transaminase (SGPT), and lactic dehydrogenase (LDH) were evaluated in three patients. Two of these showed a rise in SGOT in the range of 48 to 91 units. SCPT was elevated in one patient on serial determinations, and LDH was elevated in one patient.

Cardiac Catheterization

Cardiac catheterization was performed in four patients aged 2, 3, and 4 months and 13 years. Cardiac catheterization data are shown in table 5.

The right ventricular and pulmonary artery systolic pressures were normal to slightly elevated (25 to 43 mm Hg) in the three infants and normal in the 13 year old.

Left ventricular end-diastolic pressures were at upper limits of normal to markedly

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Figure 1
Electrocardiogram of patient 11 in 1955. Precordial leads are at one-half standardization. Anterolateral infarction and left ventricular hypertrophy with strain are present.

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Elevated in all three patients in whom they were obtained (10, 25, and 28 mm Hg).

Left-to-right shunt at the pulmonary artery level was detected by oxygen studies in two patients, both of whom survived (table 6). Dye curves revealed evidence of a shunt at the pulmonary artery level in all three patients in whom this study was performed. In two of these three patients, there was no evidence of left-to-right shunt by oxygen data. Three of the four patients had cineangiographic evidence of left-to-right shunt. In one patient (no. 10) origin of the left coronary artery could not be demonstrated either by aortic or by pulmonary artery angiography.

Of the three technics used for detecting the left-to-right shunt at pulmonary artery level, the indicator-dilution curve was the most sensitive (table 6). Of the four patients studied, only those with shunts detectable by means of oxygen saturation studies survived.

Treatment

Of the 11 patients, seven received medical treatment or no treatment, and four under-

Table 5

Catheterization Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>SVC</th>
<th>RA</th>
<th>RV</th>
<th>PA</th>
<th>Ao</th>
<th>Qp/Qs</th>
<th>Pressures</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>74</td>
<td>75</td>
<td>75</td>
<td>81</td>
<td>94</td>
<td>1.5/1</td>
<td>25/3</td>
</tr>
<tr>
<td>8 a*</td>
<td>50</td>
<td>53</td>
<td>48</td>
<td>56</td>
<td>91</td>
<td>1.3/1</td>
<td>43/0</td>
</tr>
<tr>
<td>b†</td>
<td>65</td>
<td>68</td>
<td>68</td>
<td>68</td>
<td>95</td>
<td>1/1</td>
<td>22/0</td>
</tr>
<tr>
<td>9</td>
<td>67</td>
<td>68</td>
<td>66</td>
<td>68</td>
<td>93</td>
<td>1/1</td>
<td>39/4</td>
</tr>
<tr>
<td>10</td>
<td>63</td>
<td>65</td>
<td>57</td>
<td>62</td>
<td>94</td>
<td>1/1</td>
<td>32/2</td>
</tr>
</tbody>
</table>

*Preoperative data.
†One year, 3 months following ligation of anomalous left coronary artery.

Abbreviations SVC = superior vena cava; RA and RV = right atrium and ventricle; PA = pulmonary artery; Ao = aorta; Qp/Qs = pulmonary-systemic flow ratio; LV = left ventricle.

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went ligation of the anomalous left coronary artery. In all four ventricular fibrillation developed at surgery. Of the four patients treated surgically, one died at surgery; one died 16 days following surgery of systemic emboli from a left ventricular thrombus; one died 10 months following surgery of congestive heart failure and arrhythmia, and one survived and is now asymptomatic. Repeat cardiac catheterization of the survivor 1 year and 3 months following surgery revealed persistence of a left ventricular aneurysm noted prior to surgery, mild mitral regurgitation noted prior to surgery, and restrictive disease of the left ventricle (endocardial fibroelastosis). End-diastolic pressure in the left ventricle had decreased from 23 to 9 mm Hg. Hence, in this series there is a 25% survival of patients with the appropriate diagnosis and with ligation of the anomalous left coronary artery.

Of the remaining seven patients who received either medical or no therapy, one (14%) survived. The 13-year-old survivor is doing well, although he has decreased exercise tolerance.

Both patients who survived surgery (one for 10 months and the other, still living 2 years following surgery) had repeated episodes of upper respiratory infection, bronchopneumonia, wheezing, or bronchiolitis. The latter patient's condition has been stable since 4 months following surgery. The patient who survived without surgery had a similar picture during the first year of life.

### Table 6

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Oxygen</th>
<th>Indicator dilution</th>
<th>Angiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>8</td>
<td>Yes</td>
<td>Not done</td>
<td>Yes</td>
</tr>
<tr>
<td>9</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>10</td>
<td>No</td>
<td>Yes</td>
<td>No (no left coronary artery seen)</td>
</tr>
</tbody>
</table>

### Discussion

The pathologic and clinical features, electrocardiograms, and x-rays of the patients in our series do not differ significantly from those reported in the literature.5–15 Of the 50 electrocardiograms reviewed from reports in the literature, Wesselhoeft and associates15 found anomalous left coronary artery from the pulmonary artery to be associated with myocardial infarction in 80%. All of our patients had evidence of infarction, although one had only loss of R-wave voltage when first seen. T-wave changes in the precordial leads were noted in all cases in the literature as in our series. S-T segment elevation was present in 40% of the cases in the literature and in 63% of our cases.

While there is consensus that medical treatment is indicated for congestive heart failure, there is disagreement concerning the most propitious time for surgery and the type of surgery that should be performed. Currently two types of surgical approaches, namely ligation of the anomalous left coronary artery near its site of origin and anastomosis of the left coronary artery to the aorta, are proposed.15–29

The major criterion for surgery has generally been demonstration of the anomaly. Demonstration of left-to-right shunt into the pulmonary artery, usually by angiograms, is the major criterion required for ligation. The major criterion for anastomosis is a vessel large enough technically to accomplish the procedure.20–23

In a recent paper, Wesselhoeft and associates15 proposed classifying anomalous left coronary arteries into four groups according to the clinical presentation as follows: group I, infant syndrome onset at 2 to 4 months with angina-like symptoms and cardiomegaly; group II, mitral insufficiency of undetermined etiology; group III, continuous murmur syndrome; and group IV, sudden death in adults.

All of the patients in our series fall into group I.

Of the 105 reported cases of group I infants collected from the literature by Wesselhoeft,
has not been reported in a patient older than 2 years of age.

Because both of our patients with left-to-right shunts detectable at the pulmonary artery level by oxygen studies survived, one with surgery and the other without it, we decided to examine the reports of others as to their experience with left-to-right shunts of this magnitude. In reviewing the literature, we included only patients who had cardiac catheterization performed and on whom oxygen saturation studies were reported. Because of our experience mentioned above, we divided the reported infants and children into three groups on the basis of the catheterization data as follows: group A, those with sufficient intercoronary anastomoses to be associated with an increase in oxygen saturation in the pulmonary artery at the time of cardiac catheterization; group B, those with intercoronary anastomoses insufficient to produce an increase in oxygen saturation in the pulmonary artery at the time of cardiac catheterization, but with a left-to-right shunt at the pulmonary artery level which may be detected by dye-dilution curves or by cineangiography; and group C, those with no evidence of left-to-right shunt in the pulmonary artery by any means or pulmonary artery hypertension, or both.

Although approximately 160 cases have been reported, we found only 44 cases, including four of our own (cases 8 through 11), in which cardiac catheterization had been performed in infancy or childhood and in which oxygen data and cineangiographic data, and, in some of the cases dye-dilution curves, were available (table 7). Of the 18 group A patients, all survived regardless of type of surgery or if no surgery was performed.

Of the 22 group B patients, two underwent anastomosis of the left coronary artery to the aorta and both lived. In 11 the left coronary artery was ligated and six (55%) survived. Of nine not undergoing surgery, 45% survived.

Of the four group C patients two expired when the anomalous left coronary artery was ligated, one died without surgical treatment,
and the fourth died during induction of anesthesia before surgery.

Total mortality in these 44 cases was 32%. The lower mortality rate suggests that this group is different from the total population of patients with this anomaly. These differences might be related to severity of disease or to treatment received. If we stratify these 44 patients according to age at the time of catheterization as an indicator of severity of illness, we find that group A patients were the oldest and had less severe disease (table 8). The difference between mean age of group A and group B patients is statistically significant \((P = 0.05)\). Of the group B patients, those undergoing anastomosis of left coronary artery to aorta were older than those having ligation of left coronary artery and also had less severe disease. Of those undergoing ligation, the survivors were older at the time of catheterization than the nonsurvivors. Of the untreated patients, the survivors were catheterized later and obviously had less severe disease than the nonsurvivors.

Hence, whether the improved mortality statistics on these 44 patients (32%) as compared to infants and children reported in the literature (86%) are related to the treatment the patient received or to the fact that these patients had less severe disease and lived long enough to be thoroughly studied cannot be answered at this time. The data suggest some degree of natural selection in all groups, making accurate appraisal of the merits of various therapeutic approaches difficult. Certainly, it appears that those with left-to-right shunts detectable with oxygen studies (group A) have abundant intercoronary anastomoses and will survive regardless of the type of treatment. In this group, surgery should probably be postponed until the child is old enough to have an anastomosis.

Treatment of those with angiographic or indicator-dilution evidence of left-to-right shunt but without oxygen saturation confirmation (group B) is associated with higher mortality rates regardless of whether treatment is medical or surgical. Although most will agree that a surgical approach should be attempted in the group B infant who is deteriorating, data available in the literature fail to show a significant difference in the mortality rates of infants treated either medically or surgically. In this group perhaps more consideration should be given to the possibility of performing delayed anastomosis or ligation with intensive medical therapy being the only treatment during the first year of life.

Factors which indicate the members of group B who have or will develop intercoronary collaterals and survive are not clear at the present time. Augustsson and associates\(^1\) postulate that adequacy of intercoronary anastomoses either is or is not present at birth. They are of the opinion that development of collateral circulation is not an important feature. Others write that a gradual transition may occur in functional pattern as intercoronary anastomoses enlarge.\(^2\,\(^3\) Hence, those who survive either are born with or can rapidly develop adequate intercoronary anastomoses.\(^2\)

Group C patients with inadequate intercoronary anastomoses will require either an early anastomosis or an arterial shunt into the left coronary artery.

In the future each patient with this anomaly should be evaluated with complete cardiac catheterization. These findings should be

### Table 8

<table>
<thead>
<tr>
<th></th>
<th>Group A (18 patients)</th>
<th>Group B (22 patients)</th>
<th>Group C (8 patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Survivors</td>
<td>Dead</td>
<td>Survivors</td>
</tr>
<tr>
<td>Mean Age</td>
<td>6.1 yr</td>
<td>None</td>
<td>4.5 yr</td>
</tr>
<tr>
<td>Range</td>
<td>1/3-16 yr</td>
<td></td>
<td>4-5 yr</td>
</tr>
</tbody>
</table>

**Age at Time of Catheterization: Reported Cases of Anomalous Left Coronary Artery**
correlated with clinical and other laboratory data to determine features which might allow prediction of those group B infants who will survive. For example, two of our patients with serial electrocardiographic evidence of additional myocardial infarction while on medical therapy did not survive; serial electrocardiographic changes might be useful in predicting an unfavorable outcome. The likelihood of survival without surgery as compared to the actual survival rate with specific surgical techniques should be compared. Only in this way can sufficient data be accumulated to evaluate adequately all types of surgical approaches.

At present, the younger the patient at the time of diagnosis, the less likely he is to survive. Since nearly all patients have developed ventricular fibrillation with surgery, administration of beta-adrenergic blocking agents immediately prior to surgery should be considered. The likelihood of complete cure in patients with electrocardiographic evidence of infarction is remote. All reported living patients have some residual disease.

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