Anomalous Left Coronary Artery Originating From the Pulmonary Artery

Report on 15 Cases

By Joseph Askenazi, M.D. and Alexander S. Nadas, M.D.

SUMMARY

Fifteen infants and children with the diagnosis of anomalous left coronary artery from the pulmonary trunk have been encountered at the Children’s Hospital Medical Center, Boston, Massachusetts from 1958 to 1973. After thorough clinical and laboratory evaluation, they have been treated by anticongestive measures. Nine patients have had ligation of the anomalous left coronary artery at its entrance into the pulmonary artery; one patient has undergone coronary bypass surgery.

The electrocardiogram proved to be the most helpful diagnostic clinical laboratory test. Vectorcardiograms are valuable not only in diagnosis but also in the follow-up of the patients from the prognostic point of view.

The most sensitive tool for the definitive diagnosis is an aortic root angiogram; we have no false negatives or false positives with this method.

The twelve patients with complete cardiac catheterization data could be divided into three groups, according to the pressure and magnitude of the left-to-right shunt at the pulmonary level.

All patients with an appreciable left-to-right shunt at the pulmonary level died. Patients in whom no left-to-right shunt could be demonstrated by angiography died. Half of the patients with only small left-to-right shunt survived. The results of surgical and medical treatment were identical within the three groups.

Medical management in infancy, according to coronary care principles, with definitive surgical correction at a later age, is the preferred treatment. Ligation of the anomalous left coronary artery is recommended in severely symptomatic infants with documented left-to-right shunt at the pulmonary artery level, who do not respond to medical management.

Additional Indexing Words:

Bypass surgery
Cardiac catheterization

Anomalous origin of the left coronary artery from the pulmonary trunk is a rare congenital cardiac anomaly accounting for 0.46% of Gasul’s patients with congenital heart disease and 0.26% of patients with congenital heart disease catheterized at the Children’s Hospital Medical Center in Boston. For the past sixteen years we have seen approximately one new patient per year with this anomaly in our institution.

Up to the present time, some two hundred cases have been reported, the majority having been diagnosed in infancy. The anomaly has serious prognosis with a mortality of 80–85% in the infantile group. Surgical palliation or treatment thus is highly desirable.

The object of this paper is to review our experience with this entity at the Children’s Hospital Medical Center of Boston and to propose indications for surgical intervention.

Materials

The files of our cardiac pathology registry as well as those of the cardiac catheterization laboratory and the diagnostic files of the Cardiology Department of the Children’s Hospital Medical Center for the last 16 years were reviewed. Since 1958, fifteen patients (10 males and 5 females) have been seen at the Children’s Hospital Medical Center with anomalous left coronary artery (table 1). Three of our patients (cases 1, 2 and 9 in table 1) were previously reported. The ages of our patients at the onset of symptoms ranged from 2 weeks to 6 years; 12 were infants under 6 months of age, one was 4½ years old and the other two 6 years of age. Seven had birth weights below the 15th percentile. All pregnancies were unremarkable except for two, one with vaginal bleeding at 3½ and 6½ months and the other with placenta previa and uncomplicated delivery. Eight patients underwent ligation of the anomalous left coronary artery in our institution. One additional patient had coronary artery ligation in California and one had a graft.
Table 1
Anomalous Left Coronary Artery From Pulmonary Artery at the Children's Hospital Medical Center, Boston, 1958-73

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age at onset of symptoms</th>
<th>Age at first visit</th>
<th>Chest X-ray</th>
<th>ECG</th>
<th>VCG</th>
<th>Catheterization</th>
<th>Surgery</th>
<th>Age (yrs)</th>
<th>Course</th>
<th>Postmortem</th>
</tr>
</thead>
<tbody>
<tr>
<td>D.R.</td>
<td>F</td>
<td>2 wk</td>
<td>18 day</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Ligation</td>
<td>5/52</td>
<td>Died 30 mo after surgery in CHF</td>
<td>+</td>
</tr>
<tr>
<td>A.J.</td>
<td>M</td>
<td>1 mo</td>
<td>5 wk</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Ligation</td>
<td>6/52</td>
<td>Living and well</td>
<td></td>
</tr>
<tr>
<td>G.K.</td>
<td>M</td>
<td>7 wk</td>
<td>7 wk</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>++</td>
<td>None</td>
<td></td>
<td>CHF, sudden death at 1 yr</td>
<td>+</td>
</tr>
<tr>
<td>A.L.</td>
<td>M</td>
<td>7.5 wk</td>
<td>8 wk</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Ligation</td>
<td>2/12</td>
<td>Died 24 hr postop</td>
<td>+</td>
</tr>
<tr>
<td>S.D.</td>
<td>M</td>
<td>2 mo</td>
<td>7.8 yr</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>++</td>
<td>Poudrage</td>
<td>7/12</td>
<td>Living, asymptomatic with cardiomegaly and MR</td>
<td>+</td>
</tr>
<tr>
<td>R.P.</td>
<td>M</td>
<td>2 mo</td>
<td>2 mo</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>Ligation</td>
<td>3/12</td>
<td>Died 1 hr postop</td>
<td>+</td>
</tr>
<tr>
<td>J.C.</td>
<td>M</td>
<td>2.5 mo</td>
<td>3 mo</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Ligation</td>
<td></td>
<td>Died at 5½ yrs after syncopal attack</td>
<td>−</td>
</tr>
<tr>
<td>T.F.</td>
<td>F</td>
<td>2.5 mo</td>
<td>3.5 mo</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Ligation</td>
<td>4/12</td>
<td>Living and well with mild MR</td>
<td></td>
</tr>
<tr>
<td>M.R.</td>
<td>F</td>
<td>3 mo</td>
<td>6 mo</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Ligation</td>
<td>6/12</td>
<td>Died 5 hr postop</td>
<td>+</td>
</tr>
<tr>
<td>D.P.</td>
<td>M</td>
<td>3 mo</td>
<td>3 mo</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>Thoracotomy for L bronchus obstruction</td>
<td>3/12</td>
<td>Died 2 hr postop</td>
<td>+</td>
</tr>
<tr>
<td>J.W.</td>
<td>M</td>
<td>3 mo</td>
<td>5 mo</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Ligation</td>
<td>6/12</td>
<td>CHF, died suddenly</td>
<td>+</td>
</tr>
<tr>
<td>J.B.</td>
<td>F</td>
<td>5 mo</td>
<td>8 mo</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Ligation</td>
<td>10/12</td>
<td>6/12 mo postop</td>
<td>+</td>
</tr>
<tr>
<td>K.K.</td>
<td>F</td>
<td>2 yr</td>
<td>4.5 yr</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Ligation</td>
<td></td>
<td>CHF, severe MR, sudden death 18 ½ yr</td>
<td>+</td>
</tr>
<tr>
<td>A.G.</td>
<td>M</td>
<td>6 yr</td>
<td>6 yr</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td>Living with cardiomegaly, asymptomatic</td>
<td></td>
</tr>
<tr>
<td>M.Y.</td>
<td>M</td>
<td>6 yr</td>
<td>10 yr</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Anastomosis</td>
<td>13/6/12</td>
<td>Living and well 20 mo postop</td>
<td>+</td>
</tr>
</tbody>
</table>

*Test done during follow-up.
†Done 13½ yrs postop.

Abbreviations: CHF = congestive heart failure; MR = mitral regurgitation.
anastomosis to the ascending aorta at the Massachusetts General Hospital.* Postmortem examination was performed on eight patients; seven of these hearts were available for examination. In the eighth one, whose autopsy was performed elsewhere, only the pathology report was available.

Results

Admissions Symptoms, Signs, and Laboratory Findings

History and Physical Examination

The classical symptoms of infantile angina, as described by Bland, White and Garland in 1933, were the presenting complaint in only four infants; three of these also had congestive heart failure. The other eight presented with left-sided failure (tachypnea, cough, wheezing, even pulmonary edema without angina). Among the three older children, one presented with angina, one with a murmur of mitral regurgitation and left-sided failure and the third with cardiac enlargement of unknown origin. The physical findings, detailed in 15 patients, included evidences of congestive failure (tachypnea, dyspnea, rales, hepatomegaly and cardiac enlargement) in 11 and underdevelopment (under the 3rd percentile of the Boston development charts) in ten patients. Symptoms and signs on admission are presented in figure 1. A systolic murmur was noted in 13 patients at the time of admission. Seven of these were typical mitral regurgitation murmurs, five patients had faint apical systolic murmurs of undefined quality and one infant had a lower left sternal border pansystolic murmur suggesting a ventricular septal defect. One patient, first seen at 8 months, and now 4½ years postoperative has never had any murmurs described. In addition to an apical systolic murmur, one child had on admission an apical diastolic rumble as well. No continuous murmur was heard in any patient on admission.

Enzymes

Admission SGOT and SGPT levels were evaluated in five patients all under one year of age; three showed elevated SGOT (range 75–139 u) and one an elevated SGPT (80 u).

Radiology

Chest X-rays were obtained in all 15 patients and all but patient 5 were fluoroscoped. Cardiomegaly and pulmonary venous congestion were the outstanding features in 11 infants (average cardiothoracic ratio 64%, range 59–70%) (fig. 2). The enlargement commonly involved the left ventricle; in five instances there was fluoroscopic evidence of left atrial enlargement as well. The dominant radiographic finding in patient 10 was a "white-out of the left hemithorax." Fluoroscopy revealed atelectasis with left bronchial obstruction probably due to left atrial enlargement. This is the one patient in whom no complete cardiac evaluation was available preoperatively. There was one infant with a cardiothoracic ratio of 54% and questionable pulmonary vascular engorgement.

Among the older children, one had a normal sized heart and two had modest cardiomegaly with an average cardiothoracic ratio of less than 60%.

*Personal communication, Dr. M. J. Buckley.
Electrocardiograms

The electrocardiogram, available in all but patient 10, was abnormal in every instance, as may be seen in figure 3. Left ventricular hypertrophy12 was present in all patients. In addition, right ventricular hypertrophy was noted in the patient with ventricular septal defect and pulmonary artery hypertension. Among the ten patients with superior frontal plane axis, five had left anterior hemiblock.13 The diagnosis of anterolateral myocardial infarction could be established on the basis of a Q-T pattern in 13 of 14 patients with adequate electrocardiograms. The typical pattern of a Q wave in leads I, aVL and V4-V6 was present in only five patients but 13 had significant Q and T wave abnormalities in at least one of the left ventricular leads. Only patient 3 with biventricular hypertrophy, a ventricular septal defect and pulmonary artery hypertension did not have the typical findings of myocardial infarction (fig. 4A). A typical electrocardiogram from patient 6 is shown in figure 4B.

Vectorcardiograms

Frank vectorcardiograms were obtained in eight patients on admission. The mean QRS duration was slightly prolonged (85 msec). Maximal spatial vectors to the left were increased (posteriorly and superiorly directed) with an average of 2.5 mV, indicating left ventricular hypertrophy. The loop was predominantly superior in the frontal plane with counter-clockwise inscription in six patients (fig. 5). There was one patient with a figure-of-eight loop with initial forces directed superiorly and counter-clockwise and another with a clockwise superior loop. The horizontal plane was uniformly characteristic of anterolateral myocardial infarction with clockwise rotation, the initial vectors pointing anteriorly and rightward, and the loops then progressing posteriorly.

Cardiac Catheterization

Cardiac catheterization was performed at first admission in our institution on all but patient 5. One additional child (patient 2) had a single cardiac catheterization done 13½ years after surgery. Eight patients were six months old or less at the time of the first study, one was nine months old and three children were 6, 9, and 10 years old. The clinical diagnosis was confirmed in seven of 12 patients. Cardiac catheterization data in 13 patients, including the one (patient 2) who only had a postoperative study, are summarized in table 2. Pulmonary artery systolic pressures were moderately elevated (33–55 mm Hg) in four patients (cases 1, 3, 7 and 11) including the one with the large ventricular septal defect. Mean pulmonary capillary wedge pressure was generally elevated with a maximum of 13 mm Hg; it was less than 8 mm Hg in three patients (cases 2, 8, 15). Left ventricular end-diastolic pressures were recorded in eight patients and were significantly elevated (range 10–22 mm Hg) in six.

A significant left-to-right shunt by oxygen saturation, at the pulmonary artery level, could be demonstrated preoperatively in only three instances (cases 8, 12, 14). Patient 2 had an oxygen step-up at the pulmonary artery level postoperatively. Fiberoptic oxygen study was performed in four patients (cases 7, 11, 12, 15); wide phasic oxygen variations above the pulmonary valve, sharply disappearing in the right ventricle, were shown in three of these including two without significant changes in oxygen saturation. There were no false negatives among the patients with the fiberoptic study (table 3). The presence of a left-to-right shunt was also explored by hydrogen inhalation study and by indicator dye injection into the aortic root with sampling in the main pulmonary artery. In three of seven patients, a left-to-right shunt could not be demonstrated by either of these methods. Additional left-to-right shunts were demonstrated in two patients, one at the atrial and the other at the ventricular level.

An accurate anatomic diagnosis could be made by left ventricular or supravalvar angiography in all ten patients in whom such a procedure was performed (fig. 6). The right coronary artery appeared dilated and tortuous; a small left coronary artery filled in retrograde fashion via collaterals and the main pulmonary artery actually was opacified through the anomalous left coronary artery in the six patients with the largest collaterals. Main pulmonary artery angiography was performed in three patients. In one, the left coronary artery filled from the main pulmonary artery, and in the second patient an outpouching of the main pulmonary artery suggested anomalous origin of the left pulmonary artery. Left
ventriculograms revealed a dilated chamber with poor contractions and with ejection fractions ranging from 32–66%. In eight of 11 patients ejection fractions were less than 44%. Variable degrees of mitral incompetence, as demonstrated by left atrial filling without premature ventricular contractions, were present in four of seven patients.

Treatment and Clinical Course

All patients in congestive heart failure were treated with digitalis and diuretics. Most of them were sedated with barbiturates and occasionally with morphine.

Ten patients have had surgery (table 1). One 13-year-old boy had ligation and division of the left coronary artery at its entrance into the pulmonary artery and a saphenous graft anastomosis to the aorta. Twenty months postoperatively he is doing well; neither repeat cardiac catheterization nor any arteriogram has been performed. Nine patients had only ligation of the anomalous left coronary artery; eight of these were infants with a median age of 3 months (5 weeks to 10 months). The ninth was operated upon at 5½ years having had previous poudrage of the pericardium at 7 months.

Three of the eight operated infants died within 24 hours following surgery. None of these patients fulfilled our published criteria for operation; in none was there a left-to-right shunt demonstrated preoperatively. Of the five infants who survived operation, one (who at the time of surgery had a necrotic and calcified left ventricle) continued to be in congestive heart failure and died 30 months postoperatively. Catheterization in this child 20 months postoperatively revealed a severely dilated left ventricle (end-diastolic volume index of 194 ml/m²) and impaired ventricular function with an ejection fraction of 0.25. A second patient (case 11) died suddenly six months after coronary artery ligation in congestive heart failure due to papillary muscle dysfunction.

The last three patients are presently asymptomatic 4½, 5, and 13½ years postoperatively. One of them (case 8), a girl with residual mitral regurgitation, is on digitalis and has a strain pattern in the electrocardiogram. Chest X-ray shows her heart still to be enlarged, but much smaller than preoperatively. The second one (case 2), a 14-year-old boy, is asymptomatic with cardiomegaly, left ventricular strain pattern, and a continuous murmur. Repeat cardiac catheterization 13½ years postoperatively revealed recanalization of his surgical occlusion with shunting from the right coronary artery to the left coronary artery and to the pulmonary artery. He is now awaiting coronary artery bypass surgery. The third

Figure 4

A) ECG taken from patient 3 showing left atrial enlargement and biventricular hypertrophy. He also had a ventricular septal defect and pulmonary artery hypertension. B) ECG taken from patient 6 at two months of age demonstrating the typical electrocardiographic findings in this anomaly. Note the acute current injury in the anterolateral wall.
one (case 12) is a girl with no significant murmur and only slight cardiac enlargement. She is the only patient who has a normal electrocardiogram.

Five patients were treated medically. Only one is alive (case 14), a 12½-year-old boy who is asymptomatic and the parents refuse surgery in spite of the very severe electrocardiographic changes and cardiomegaly. Two patients, one an 18-year-old girl (case 13), the other a 5½-year-old boy (case 7), died after a syncopal episode. The fourth patient with associated large ventricular septal defect died at one year in chronic congestive heart failure. The fifth patient died suddenly at three months with evidence of myocardial infarction and pericardial tamponade (case 10).

Because of the long follow-up, patient 13 is of unusual interest. A brief summary of her course follows.

K. K., a 4½-year-old asymptomatic girl, was first seen at the Children's Hospital Medical Center for evaluation of a heart murmur in 1956. The murmur was first noted at 2 years of age. Physical examination on her first visit revealed a well developed, well nourished girl with a grade III/VI blowing apical systolic murmur radiating to the axilla; a loud mid-diastolic rumble was also heard at the apex. Chest X-ray showed a markedly enlarged heart with over-all enlargement of both ventricles, considerable left atrial enlargement and slightly increased pulmonary markings. Electrocardiogram (fig. 7A) disclosed a superior frontal plane axis (−40°), left ventricular hypertrophy and Q-T pattern of old anterolateral myocardial infarction. The patient was put on penicillin prophylaxis and was followed with the diagnosis of mitral regurgitation of unknown origin. During the fifth year of her life she was hospitalized elsewhere three times for recurrent bronchitis and pneumonitis. At 6 years of age, cardiac catheterization was performed (table 2) confirming the impression of mitral regurgitation and probable myopathy; the significance of a 5–8% increase in oxygen saturation in the main pulmonary artery was not appreciated. Subsequently the patient was treated with digoxin because of her cardiac enlargement. Chest X-rays continued to show cardiomegaly with left atrial enlargement and increasing passive congestion. Electrocardiograms showed left atrial enlargement, inverted T waves over V₅, aVL, and V₆, and decreased R wave amplitude in V₄. Repeat cardiac catheterization at 7 years of age showed a left ventricular end-diastolic pressure of 22 mm Hg. The patient continued to do well until 13 years of age when dyspnea on exertion and mild exercise intolerance developed.

At fifteen years of age she was again admitted to the Children's Hospital. Vectorcardiogram at that time showed counter-clockwise rotation in the frontal plane with left anterior hemiblock, clockwise rotation of horizontal loop with decreased anterolateral forces consistent with anterolateral myocardial infarction. The left ventricular and

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**Figure 5**

Vectorcardiogram taken from patient 11 showing abnormal clockwise rotation of the horizontal plane (H) projection and counter-clockwise inscription of the frontal plane (F), characteristic of anterolateral myocardial infarction.
Catheterization and Angiography

Cardiac Catheterization and Angiography Results in 13 Patients with Anomalous Left Coronary Artery

<table>
<thead>
<tr>
<th>Patient</th>
<th>Catheterization age</th>
<th>MPA</th>
<th>PCW</th>
<th>LV</th>
<th>SA</th>
<th>RA</th>
<th>MPA</th>
<th>SA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. D.B.</td>
<td>3 wks &amp; 1 mo</td>
<td>40/19</td>
<td>15/8 (8)*</td>
<td>—</td>
<td>112/86 (90)</td>
<td>87</td>
<td>84</td>
<td>96</td>
</tr>
<tr>
<td>2. A.J.</td>
<td>14 yrs‡</td>
<td>17/8 (13)</td>
<td>11/8 (7)</td>
<td>95/10</td>
<td>95/65 (75)</td>
<td>79</td>
<td>84</td>
<td>96</td>
</tr>
<tr>
<td>3. G.K.</td>
<td>2 mos</td>
<td>55/18 (37)</td>
<td>10/15 (12)</td>
<td>—</td>
<td>80/37</td>
<td>65</td>
<td>80</td>
<td>95</td>
</tr>
<tr>
<td>4. A.L.</td>
<td>4 wks</td>
<td>20/18</td>
<td>—</td>
<td>—</td>
<td>90/42 (55)</td>
<td>60</td>
<td>62</td>
<td>92</td>
</tr>
<tr>
<td>5. D.S.</td>
<td>2 mos</td>
<td>20/13</td>
<td>—</td>
<td>—</td>
<td>80/40</td>
<td>67</td>
<td>70</td>
<td>98</td>
</tr>
<tr>
<td>7. J.C.</td>
<td>3 mos</td>
<td>33/11 (16)</td>
<td>14/19 (13)</td>
<td>—</td>
<td>84/46</td>
<td>67</td>
<td>67</td>
<td>99</td>
</tr>
<tr>
<td>8. T.F.</td>
<td>4 mos</td>
<td>22/9 (13)</td>
<td>9/10 (7)</td>
<td>80/9</td>
<td>83/47 (70)</td>
<td>64</td>
<td>69</td>
<td>97</td>
</tr>
<tr>
<td>9. M.R.</td>
<td>6 mos</td>
<td>27/12 (14)</td>
<td>12/14 (9)</td>
<td>—</td>
<td>—</td>
<td>65</td>
<td>65</td>
<td>84</td>
</tr>
<tr>
<td>11. J.W.</td>
<td>6 mos</td>
<td>38/12 (22)</td>
<td>18/14 (12)</td>
<td>82/9-15</td>
<td>92/64 (74)</td>
<td>60</td>
<td>59</td>
<td>95</td>
</tr>
<tr>
<td>12. J.B.</td>
<td>9 mos</td>
<td>28/11 (19)</td>
<td>12/13 (11)</td>
<td>100/15</td>
<td>100/60 (80)</td>
<td>69</td>
<td>77</td>
<td>96</td>
</tr>
<tr>
<td>13. K.K.</td>
<td>6 yrs</td>
<td>28/13 (18)</td>
<td>/19 (14)</td>
<td>—</td>
<td>92/55 (65)</td>
<td>73</td>
<td>76</td>
<td>97</td>
</tr>
<tr>
<td>14. A.G.</td>
<td>9 yrs</td>
<td>18/11 (16)</td>
<td>11/13 (9)</td>
<td>95/9</td>
<td>95/65 (81)</td>
<td>76</td>
<td>81</td>
<td>98</td>
</tr>
<tr>
<td>15. M.Y.</td>
<td>9½ yrs</td>
<td>17/10 (13)</td>
<td>8/9 (7)</td>
<td>112/10</td>
<td>105/74 (95)</td>
<td>78</td>
<td>79</td>
<td>97</td>
</tr>
</tbody>
</table>

*Mean pressures in parentheses. 
†Left atrial pressure. 
‡Postoperative study.

Abbreviations: MPA = main pulmonary artery; PCW = pulmonary capillary wedge; SA = systemic artery; RA = right atrium; L-R = left-to-right shunt; R-L = right-to-left shunt; ASC = ascending aorta; LV = left ventricle; ASD = atrial septal defect; VSD = ventricular septal defect; MR = mitral regurgitation; PS = pulmonary stenosis; PV = pulmonary vein; DR = diastolic rise in saturation; Y = yes; N = no; A = aorta; Sugg = suggestive.

Aortic root angiograms revealed severe mitral regurgitation, dilatation and poor contraction of the left ventricle, marked enlargement of the right coronary artery with numerous large collaterals filling, in retrograde fashion, the left coronary artery to the pulmonary artery. The diagnosis of mitral regurgitation secondary to anomalous left coronary artery was clear, but because of her relatively mild symptomatology and the serious nature of the proposed surgical intervention, including mitral valve replacement as well as transplantation of the coronary artery, it was decided to continue to treat her conservatively.

On her last visit six months prior to her death, symptoms and physical findings had not changed but the electrocardiogram (fig. 7C) showed a decrease in R waves over the precordium and the appearance of deep Q waves in V4. At age 18½ years, during a ballet lesson, she developed sudden ventricular fibrillation responding to external cardiac massage, xylocaine and countershock; she remained unconscious and died several hours later in intractable pulmonary edema.

This tragic course of events may be excused only by our relative inexperience in the 1950s and 60s with surgical and diagnostic techniques that are commonplace in 1974.

Electrocardiographic changes of interest during the clinical follow-up period include changes in mean frontal plane axis in patients 2, 5 and 8. The first two developed a superior axis postoperatively whereas in patient 8, preoperative left axis deviation disappeared after coronary artery ligation. The Q waves also tended to diminish in amplitude and width in the survivors of surgery and in some of them the electrocardiograms were indistinguishable from those characteristic of cardiomyopathies or endocardial fibroelastosis.

In all patients in whom vectorcardiograms were obtained typical patterns of anterolateral infarction were seen. General improvement with time, or following ligation of the anomalous left coronary artery, resulted in improvement of the appearance of the loop in the horizontal plane, as pointed out by Ellison and Restieux. Persistent clockwise rotation in the horizontal plane reflects a large area of fibrosis, lack of
revitalization and high incidence of dyskinesia as well as asynergy of the anterior and apical segments suggested by Gorlin (personal communication). In

Table 3
Presence of Shunt by Various Methods in 12 Patients with Anomalous Left Coronary Artery

<table>
<thead>
<tr>
<th>Patient</th>
</tr>
</thead>
</table>
| 1. D.B. (a) | + 0 0 0 0 0
(b) | - 0 - - - +
(c) | - 0 0 0 0 -
| 2. A.J. | + 0 0 0 0 +
| 3. G.K. | + 0 0 0 0 + R-L
| 4. A.L. | - 0 0 0 0 -
| 5. D.S. | - 0 0 0 0 +
| 7. J.C. (a) | - - 0 0 -
(b) | - 0 0 0 0 +
| 8. T.F. | + 0 0 0 0 +
| 9. M.R. | - 0 - 0 -
| 11. J.W. | - + 0 0 + +
| 12. J.B. | + + 0 0 + +
| 13. K.K. | 0 0 0 0 +
| 14. A.G. | + 0 0 0 0 +
| 15. M.Y. | + + 0 0 + +

Abbreviations: FO = fiberoptic; H+ = hydrogen; AA = ascorbic acid; CG = cardiogreen; - = negative; + = positive; 0 = not done.

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creased cardiothoracic ratio in the X-rays, particularly associated with mitral regurgitation, was of serious significance. All three patients with cardiothoracic ratio of 0.65 or greater died. All four of the postoperative patients with no significant cardiac enlargement are still alive and well.

Pathology

The anomalous left coronary artery arose from the left posterior sinus in all eight specimens available for examination. The right coronary artery was dilated and tortuous in every instance; anastomotic channels with the left coronary artery have been demonstrated by barium sulfate injection into the right coronary artery. The hearts were enlarged with a weight of 1.5 to 2.5 times normal. The left ventricle was uniformly dilated with variable degrees of endocardial fibroelastosis and fibrosis with atrophy of the anterolateral papillary muscle. Old and recent myocardial infarctions and diffuse myocardial fibrosis, apical scarring and aneurysmal dilatation with calcification were present in two specimens. The chordae tendineae were thickened, shortened and the edges of the anterior leaflet of the mitral valve were rolled and thickened. Patient 3 had a ventricular septal defect with a quadro-cusp aortic valve. Two
patients had renal anomalies: patient 4 had a horse-shoe kidney and patient 13 had a double collecting system in the left kidney. Patient 10 had a bronchomalacia of the left main bronchus.

Discussion

Anomalous left coronary artery arising from the pulmonary trunk was first described by Brooks in two adults at necropsy in 1886. In 1911 Abrikossoff reported on an autopsy on a 5 month old baby. In 1933 Bland, White and Garland reported the clinical description and demonstrated the electrocardiographic abnormalities in a three month old baby. Since then, nearly 200 cases have been reported. Accounts dealing with pathophysiology, hemodynamics and surgical treatment were given by Edwards, Augustson, Nadas, Cooley, Wesselhoeft and Perry and Scott. Edwards proposed that in every patient there is a
Serial electrocardiograms taken from patient 13. A) At 4½ years of age, left axis deviation with Q-T pattern of old anterolateral myocardial infarction. B) At 15 years of age it shows diminished R wave amplitude over the precordium with ST-segment elevation in V6. C) Half year prior to her death it shows further decrease in precordial R wave voltage and appearance of new deep Q wave in V6.

gradually changing functional pattern of the coronary flow. In the first phase (newborn period) the anomalous left coronary artery is supplied by the relatively high pulmonary artery pressure. In the second phase, after establishment of collateral vessels, the anomalous left coronary artery carries blood into the pulmonary artery (adult type). He assumed that during the transitional phase (beyond the neonatal period), corresponding to the time of onset of symptoms of myocardial ischemia, the flow of blood into the part of the myocardium supplied through the anomalous left coronary artery might be at an all time low. Survival would then depend upon the extent of collateral vessel development and the predominance of the right coronary artery. In a fourth phase, proposed recently, anastomoses no longer function as “feeding” vessels but rather act as a “steal” for the drainage system. Interestingly, Brooks in 1886 had considered that the “pulmonary coronary branch acted as the channel by which the blood was drained away.”

The clinical features of our patients were very similar to those described in the literature. In 13 of 14 (92%) the electrocardiograms showed evidence of anterolateral myocardial infarction. The only patient whose electrocardiogram did not show myocardial infarction pattern had a large ventricular septal defect with pulmonary hypertension (fig. 4A). In a similar case in the literature the electrocardiograms also disclosed right axis deviation with biventricular hypertrophy. Absence of myocardial infarction pattern may be explained by the anomalous coronary receiving adequate perfusion from the hypertensive pulmonary artery. Closure of the ventricular septal defect in this patient resulted in a drop in pulmonary artery pressure, myocardial ischemia and death.

Cardiac catheterization is fraught with more than usual danger in these patients. Among our 13 patients (catheterized 18 times) two (patients 1 and 13) experienced ventricular fibrillation three times during their study. Both were successfully cardioverted. Two deaths in Noren’s series and one among Sabiston’s cases were related to cardiac catheterization with selected coronary arteriography.

Reviewing our experience with shunt detection into and out of the pulmonary artery by various methods including oximetry, fiberoptics, hydrogen, ascorbic acid, cardiogreen, and angiography, the only positive statement one can make is that in all cases where a shunt was present, angiography was able to demonstrate it. Injection of contrast material into the aortic root visualized the pulmonary artery in all cases where other methods indicated shunt and was also positive in six patients in whom oximetry failed to reveal any left-to-right shunt. The other methods listed were not performed with enough regularity for us to be able to make a definite statement as to their sensitivity or specificity. It seems, on the basis of the experience of our 13 patients, that supravalvar aortogram is probably the most sensitive method of shunt detection.

Wesselhoeft and her associates, in a review of the literature, have found a total mortality of 89% during the first year of life among 105 patients with signs and symptoms of myocardial infarction or myopathy. Twenty of these patients were treated surgically with ten survivors (50%). Medical mortality was close to 95%. Among our nine patients with infantile syndrome treated surgically (eight with left coronary artery ligation and one with poudrage), we have five survivors beyond one year of age. This survival rate is about the same as that quoted by Wesselhoeft and others for left coronary ligation. Among the three patients with infantile syndrome treated conservatively, two died before one year of age and one survived to 5½ years at which time he died after a major syncopal attack.

Medical survival was appreciably better among a group of six infants treated by the Texas Children’s Hospital group. There were five survivors beyond one year of age and the sixth one is alive at 7 months.
All these babies had electrocardiographic patterns of myocardial infarction.

Perry and Scott reviewed four cases of their own and 40 from the literature in regard to survival in the light of treatment as well as the presence and size of the left-to-right shunt. The group they designated as group A, had wide open anastomoses from right coronary artery to left coronary artery into the pulmonary artery resulting in a sizable left-to-right shunt detectable by oximetry; all these patients survived the first year of life, irrespective of treatment. The small group of patients in category C, without any detectable shunt into the pulmonary artery, all died, irrespective of treatment. Finally, the intermediate group B had a 50% chance to survive, again irrespective of whether they were treated medically or through left coronary ligation.

Our own experience with 12 patients (table 4) supports in all respects Perry and Scott’s conclusions.

There were two children in the group published by Perry and Scott and one among our 15 who had anastomosis of the left coronary artery to the aorta and all of these survived. We re-emphasize however, that none of these were infants and also that, according to the most recent article by El-Said et al., a sizable portion of the vein grafts will clot postoperatively. There were only two of six patients restudied within months of the anastomosis where the flow of blood was unimpeded into the coronary artery. Two of the others were completely occluded and the remaining two grafts seemed to have stenosed.

On the basis of this recent review of our own patients, taking into consideration the advances in surgical treatment, we would suggest the following therapeutic plan for a patient with an anomalous left coronary artery. 1) All patients with the clinical picture of recent myocardial infarction, regardless of age, should be treated according to modern coronary care principles, combating arrhythmias and cardiogenic shock. 2) All patients with a clinical picture suggesting anterolateral myocardial infarction should be studied by experts in cardiac catheterization and angiography. 3) In infants in whom catheterization demonstrates a sizable left-to-right shunt, conservative treatment is recommended until the patient becomes a candidate for coronary bypass. If the clinical picture deteriorates, ligation of the left coronary artery is indicated. 4) In patients with a small left-to-right shunt or no shunt at all, the prognosis with conservative treatment is so poor that attempts at anastomosis, even in infants, should be attempted. As a minimal accomplishment of this procedure, the coronary artery would be separated from the pulmonary artery and optimally effective perfusion might be accomplished. 5) Patients whose dominant clinical picture is mitral regurgitation should have their mitral valve replaced in addition to appropriate coronary artery surgery, including a jump graft and if possible even aneurysmectomy.

References
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Table 4
Mortality Experience in 55 Cases of Anomalous Left Coronary Artery with Cardiac Catheterization and Oximetry Study*

<table>
<thead>
<tr>
<th>Group</th>
<th>Status</th>
<th>Anastomosis</th>
<th>Ligation</th>
<th>No surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Percent/group</td>
<td></td>
<td></td>
</tr>
<tr>
<td>A (N = 21)</td>
<td>Living</td>
<td>2</td>
<td>0</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>Dead</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>B (N = 27)</td>
<td>Living</td>
<td>2</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>Dead</td>
<td>0</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>C (N = 7)</td>
<td>Living</td>
<td>—</td>
<td>—</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Dead</td>
<td>—</td>
<td>—</td>
<td>1</td>
</tr>
</tbody>
</table>

*Forty-three of the patients in this table were collected from the literature and reported on by Perry and Scott. The other 12 patients are from the present study.
15. Brooks H St J: Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery with some remarks upon the effect of this anomaly in producing cirsoid dilatation of the vessels. J Anat Physiol 20: 26, 1885-6
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J Askenazi and A S Nadas

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