Congenital Coronary Arteriovenous Fistula
Associated with Large Patent Ductus

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Several excellent reviews have been published recently describing the anatomic and clinical findings in patients with congenital coronary arteriovenous fistulas or with communications between the coronary arteries and the cardiac chambers.1-3

The recognition of this clinical entity presents real difficulty, since it may easily be confused with other lesions causing a continuous or to-and-fro murmur, such as patent ductus arteriosus, aortopulmonary septal defect, ruptured aneurysm of an aortic sinus, interventricular septal defect with aortic insufficiency, and arteriovenous communications within the chest wall or lungs. In some instances only a systolic murmur has been recognized. For this reason both cardiac catheterization and angiocardiography are usually required for accurate diagnosis. Of the diagnostic measures available retrograde aortography affords the best opportunity for an accurate diagnosis.

We have collected 38 cases from the literature. The first case from Steinberg's paper was discarded, since the patient had an anomalous coronary artery arising from the pulmonary artery. Although functionally similar in some respects, cases with anomalous origin of coronary arteries are best placed in a separate category. The true figure may be higher, since, as Steinberg pointed out, some coronary artery aneurysms that were considered primary lesions may have resulted from arteriovenous fistulas that were not detected at autopsy.3

Analysis of the 39 cases including our own yields the following information.

In 19 cases the right coronary system was involved and the artery entered the coronary vein or sinus in 5,4-8 the right atrium in 6,2, 9-13 the right ventricle in 7,8,14-17 and the pulmonary artery in 1.1

In 16 cases the left coronary system was involved, and the artery entered the coronary vein or sinus in 4,1,18-20 the right atrium in 2,21 the left atrium in 2,22,23 the right ventricle in 4,24-27 the left ventricle in 2,28,29 and the pulmonary artery in 2.30,31

In 1 case both right and left coronary artery systems participated in the communication with the pulmonary artery.32 In 1 case both the right and left coronaries communicated through blood spaces in the myocardium with a common ventricle.33 In 3 cases information regarding the exact anatomic arrangement was not available.3,34,35

It is apparent that left heart cavities are involved infrequently and indeed the term arteriovenous fistula appears inappropriate for these communications.

Associated congenital defects, which have been encountered in 6 cases, include pulmonary atresia, pulmonary atresia with interventricular septal defect, single ventricle, and patent ductus in 2. Indeed, in our case the patent ductus was by far the more important lesion, and the associated severe pulmonary hypertension seemed chiefly responsible for the death.

Our case, although completely studied preoperatively, remained undiagnosed. This is somewhat understandable in view of the associated large patent ductus that dominated the clinical picture and presumably caused severe pulmonary hypertension.
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![Image](fig1.jpg)  
**Fig. 1.** Posteroanterior and left anterior oblique chest roentgenograms showing right atrial, right ventricular, and pulmonary artery enlargement.

**CASE REPORT**

R.C. This Negro female infant showed symptoms of cardiac failure at age 4 months. At 16 months she developed generalized edema and hepatomegaly and was digitalized. On March 19, 1956, at 18 months she was admitted to Dooley Hospital for study.

There was no evidence of cyanosis. The blood pressure was 88/48, the pulse was 120, the respirations were 80, and the weight was 16.5 pounds. The left hemithorax was prominent with a forceful apical heart beat. A thrill was noted over the left upper chest. A grade-IV systolic murmur was heard best in the third and fourth interspaces to the left of the sternum. The pulmonic second sound was considerably accentuated.

The chest roentgenograms showed considerable cardiac enlargement, particularly of the right ventricle (fig. 1). The pulmonary artery was greatly enlarged. The electrocardiogram showed right ventricular strain (fig. 2). The hematocrit value was 49 and the hemoglobin was 8.0 Gm. per cent.

Cardiac catheterization studies in March 1956 revealed a pulmonary artery pressure of 54/24 (table 1). There was an increase in oxygen saturation from the inferior vena cava to the right atrium of 20 per cent, from the low right ventricle to the high right ventricle of 13 per cent and from the low right ventricle to the pulmonary artery of 27 per cent. The tentative diagnoses were interatrial septal defect and patent ductus arteriosus. Interventricular septal defect and pulmonary insufficiency were also considered.

A retrograde brachial aortogram was advised but the family refused.

The child was readmitted to the hospital 1 year later on December 28, 1956. At that time the blood pressure was 90/60, the pulse was 140, respirations were 38, and the weight was 22 pounds. The physical findings were similar to those on the previous admission. The pulmonic second sound was perhaps more accentuated and there was a high-pitched, early diastolic murmur along the left sternal border suggesting pulmonary insufficiency. A note was made that no diastolic murmur could be heard to the right of the sternum and no mid-diastolic murmur at the apex of the heart. Occasional rales were audible at the lung bases. The liver was felt 2 cm. below the costal margin. The red cell count was 3.4 million, the hemoglobin 8.2 Gm. per cent, the blood urea nitrogen was 27 mg. The pulmonary artery, right ventricle, and possibly the right atrium were markedly enlarged on the roentgenograms. There was suggestive enlargement of the left ventricle. Increased pulsations were seen in the pulmonary vascular bed on fluoroscopy.
A retrograde aortogram was performed through the right brachial artery. The pulmonary artery and right heart both opacified at one half second and a patent ductus was thought to be visible. At one second there was definite opacification of the right atrium and hepatic veins. The pulmonary artery was obviously quite large (fig. 3).

On the basis of these findings a tentative diagnosis of large patent ductus arteriosus with pulmonary insufficiency and tricuspid insufficiency was made. The possibility of a ruptured sinus of Valsalva into the right ventricle was also considered, but the proper interpretation of the aortogram was not made.

The patient was admitted a third time for surgery on May 11, 1957. At the time of admission there was evidence of bronchopneumonia, and the temperature was 102 °F. Crepitant rales were heard in the left lower lung field. There was a grade-III systolic murmur to the left of the sternum. The pulmonic second sound was loud and snapping. The liver was down 1 fingerbreadth. The hematocrit value was 44, and the hemoglobin was 12.3 Gm. per cent. An electrocardiogram again showed right ventricular strain. Chest roentgenogram showed evidence of congestion and patchy pneumonitis in the left lower lobe.

Operation was carried out through a bilateral transsternotomy incision 17 days later, after clearing of the pneumonitis. The pump oxygenator was prepared for possible use. A large patent ductus was found. Pressure readings were as follows: ductus opened, aortic pressure 76/40, mean 58, pulmonary artery pressure 76/40, mean 58; ductus closed, aortic pressure 84/50, mean 64, pulmonary artery pressure 58/28, mean 40. After division of the ductus there remained a thrill over the right atrium. Further examination revealed a vessel roughly 0.5 cm. in diameter, branching from a dilated left coronary artery and passing through the transverse sinus behind the aorta and pulmonary artery to empty into the right atrium behind the origin of the superior vena cava (figs. 4 and 5). The anomalous coronary vessel was ligated with several sutures near its entry into the atrium. No change occurred in the electrocardiogram.

For 24 hours after surgery the postoperative course was uneventful, but then the child died rather suddenly, apparently from a cardiac arrhythmia, about 30 hours postoperatively.

At postmortem examination there was intense pulmonary congestion of both lungs. The heart after fixation weighed 115 Gm., and there was hypertrophy of both ventricles. The left ventricle measured 7 mm., 7 mm., and 6 mm. at the base, midportion, and apex, respectively. The right ventricle showed an almost compact wall of hypertrophied

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**Table 1.—Cardiac Catheterization Data (March 1956)**

<table>
<thead>
<tr>
<th></th>
<th>Pressure</th>
<th>Per cent sat.</th>
<th>Hgb.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Main pulmonary artery</td>
<td></td>
<td>86.0</td>
<td></td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>54/20</td>
<td>84.0</td>
<td></td>
</tr>
<tr>
<td>High right ventricle</td>
<td></td>
<td>70.0</td>
<td></td>
</tr>
<tr>
<td>Low right ventricle</td>
<td>56/0</td>
<td>57.0</td>
<td>6.8</td>
</tr>
<tr>
<td>Low right atrium</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td></td>
<td>37.0</td>
<td></td>
</tr>
<tr>
<td>Femoral artery</td>
<td></td>
<td>80/24</td>
<td>97.8</td>
</tr>
<tr>
<td>Left ventricular output</td>
<td></td>
<td>5300 ml./min.</td>
<td></td>
</tr>
<tr>
<td>Right ventricular output</td>
<td></td>
<td>1800 ml./min.</td>
<td></td>
</tr>
<tr>
<td>Patent ductus shunt</td>
<td></td>
<td>3500 ml./min.</td>
<td></td>
</tr>
<tr>
<td>Coronary to right atrium</td>
<td></td>
<td>600 ml./min.</td>
<td></td>
</tr>
</tbody>
</table>

*Flows computed from oxygen consumption derived from Benedict’s tables (for weight and age). Caval oxygen content based on inferior vena cava alone.*
Fig. 3. Retrograde aortogram in extreme left anterior oblique position. *Top left.* Opacification of the aortic sinuses and aneurysmal dilatation of the first portion of the left coronary artery (0 sec.). *Top right.* Region of patent ductus seen. Pulmonary artery opacified. Right atrium well filled (0.5 sec.). *Bottom left.* Hepatic veins and right ventricle filling. Main pulmonary artery clearly outlined. Ascending aorta now empty. Aneurysmal dilatation of left coronary artery identified (1.0 sec.). *Bottom right.* Right ventricle well defined in diastole (1.5 sec.).
Histologic examination of the shunt revealed a greatly thickened wall containing little muscle tissue in some portions of the wall and a great excess of elastic tissue and collagen (fig. 6). The amount of connective tissue varied throughout the wall and produced irregularly thickened areas. Histologic examination of the pulmonary vessels revealed only minimal change despite the high pulmonary arterial pressure.

**DISCUSSION**

If a congenital coronary arteriovenous fistula had been suspected, the diagnosis could probably have been made from the evidence on the retrograde aortogram. In retrospect, the aneurysmal dilatation of the left coronary is well opacified at .5, 1, and 1.5 seconds, and the various branches are clearly seen at 1 second. The opacification of the right atrium was heavy, and even the hepatic veins filled. Since the aortogram also demonstrated a patent ductus we explained the opacification of the right heart as due to pulmonary insufficiency, which was present clinically, and tricuspid insufficiency. However, such an explanation seems illogical in retrospect, in view of the strong opacification of the right atrium. The possibility of a ruptured aneurysm of the sinus of Valsalva into the right atrium was entertained and in fact was the chief reason for preparing the pump oxygenator. However, this interpretation of the aortogram involved the assumption of an aneurysm of the left coronary sinus, a very rare condition. Rupture into the right atrium would not be anticipated.36, 37

In most cases the murmur has been described as continuous, sometimes closer to the ear than one usually associates with a patent ductus. However, to-and-fro murmurs and systolic murmurs have been described. In our case the loud systolic murmur of the patent ductus possibly obscured any murmur that might have originated from the arteriovenous fistula.

The electrocardiographic findings are available in 17 cases and were normal in 8. In 5 others either coronary artery disease or unrelated cardiac defects were present and could well explain the electrocardiographic changes.

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**Fig. 4.** Aneurysmal left coronary artery showing origin of anomalous branch as observed during surgery.

**Fig. 5.** Postmortem view showing course of anomalous coronary branch with entry into right atrium.
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Right axis deviation and atrial fibrillation were found in 1 patient with congestive heart failure in whom a large shunt passed from the left coronary artery to the coronary sinus. S-T and T-wave abnormalities occurred in a patient in whom the right coronary communicated with the pulmonary artery. In the patient discussed by Edwards and Burchell\textsuperscript{37} electrocardiographic evidence of ischemia was thought to be due to the coronary arteriovenous fistula between the right coronary and right atrium. In our own case right ventricular strain can be attributed to the presence of patent ductus arteriosus and pulmonary hypertension.

Of interest is the size of the arteriovenous communication, which in the majority of reports has ranged from 1 to 3 mm. Somewhat larger fistulas ranging in diameter from 5 to 20 mm. were described by Trevor,\textsuperscript{15} Mozen,\textsuperscript{22} Baylis and Campbell,\textsuperscript{30} and Fell et al.\textsuperscript{14} In our own case the opening was found to be approximately 2 mm. Nevertheless a moderate-size shunt was present, as evidenced by aortography and by the calculation from cardiac catheterization data. From the small amount of data available it is apparent that shunts of considerable magnitude have been encountered: 2.0,\textsuperscript{1} 1.7,\textsuperscript{26} 5.2,\textsuperscript{1} 3.9,\textsuperscript{16} 0.59 L. (this paper), or expressed as per cent of left ventricular output: 29, 39, 47, 55, and 11.

We have examined all of the reported cases to determine in what number abnormally positioned coronary arteries contributed to the fistula. Brown and Burnett\textsuperscript{26} reported an accessory left coronary that arose from the aorta and entered the right ventricle at the lower border of the pulmonary conus. Björk and Crafoord\textsuperscript{31} described an abnormal artery, arising as a branch of the left coronary, which crossed the pulmonary artery and communicated with this vessel. In the case presented by Johnson\textsuperscript{13} an anomalous right coronary coursed behind the aorta and superior vena cava to enter the right atrium. In our case an anomalous branch of the left coronary artery took a similar course behind the aorta and pulmonary artery and entered the right atrium behind the superior vena cava at its junction with this chamber.

The physiologic importance of the lesion and the prognosis for these patients must vary with the volume of the arteriovenous shunt. Although a number of the patients have remained asymptomatic, others have shown significant degrees of cardiac enlargement and even congestive heart failure. Other complications include subacute bacterial endarteritis and myocardial ischemia developing in the area beyond the arteriovenous communication.\textsuperscript{2} Associated aneurysms have been present in some of the cases and in 5 of these thrombus formation was found in the aneurysm.\textsuperscript{2, 11, 12, 31, 32} Complete clotting of the anomalous vessel and aneurysm occurred in 1 of these.\textsuperscript{31} Dilatation, sometimes of aneurysmal proportion, may be expected in the proximal coronary artery.\textsuperscript{12, 14, 15, 32, 33}

In view of the numerous possibilities for complications, surgical treatment should usu-
ally be recommended. Of the 39 cases 17 have been submitted to surgery.\textsuperscript{1, 7, 8, 11-14, 22-24, 27, 31, 35} In 4 of these exploration only was performed. Ligation of the involved artery or arteries has appeared to be a curative procedure and was accomplished in all of the other cases. An aneurysm at the site of the communication may require excision or incision to allow closure of the fistulous communication with the cardiac chamber. In the occasional case the problem may appear so formidable that the surgeon may find it useful to employ extracorporeal circulation with cardiac arrest.\textsuperscript{14} Of the 13 cases in which definitive surgery was attempted all survived except the 1 reported in this paper.

**SUMMARY**

An unusual case of congenital coronary arteriovenous fistula is reported, bringing the total number of reported cases to 39.

This is the second case in which the left coronary artery system communicated directly with the right atrium, and also the second case associated with a patent ductus.

Our case is well documented by cardiac catheterization data, retrograde aortography, and postmortem findings.

Although surgical correction of both the patent ductus and coronary arteriovenous fistula was accomplished without great difficulty, the patient died 30 hours postoperatively, presumably from the effects of severe pulmonary hypertension.

**SUMMARIO IN INTERLINGUA**

Es reportate un caso inusual de congenite fistula arterio-venose coronari. Illo augmenta le numeral de tal casos reportate in le litteratura a un total de 39.

Isto es le secunde caso in que le sistema del arteria coronari sinistre communica directemente con le atrio dextere. Illo es etiam le secunde caso associate con ducto patente.

Nostre caso es ben documentate per catheterisation cardiale, aortographia retrograde, e datos necropsial.

Ben que le correction chirurgic del ducto patente e del fistula arterio-venose coronari esseva effectuate sin grande difficultates, le patiende moriva 30 horas post le operation, apparentemente como resultato de sever grados de hypertension pulmonar.

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