Diagnosis and Surgical Correction of Congenital Coronary Artery-Coronary Sinus Fistula

By J. Alex Haller, Jr., M.D., and Joseph A. Little, M.D.

Congenital coronary arteriovenous fistula may closely simulate a patent ductus arteriosus. For this reason it is an important lesion that should be considered in the evaluation of a child with a continuous murmur near the base of the heart. The following case report illustrates the confusion that this abnormality may produce and outlines the diagnostic tests that eventually led to the correct diagnosis. In addition, this patient is apparently the first to have surgical correction of a proved right coronary artery-coronary sinus fistula.

Case Report

L.S.T., C.H. no. 74449, was first seen at the age of 5 years in the itinerant heart clinic for children in the Henderson County Health Department in February 1958. The patient was referred to the clinic by a pediatrician who had noted that the child had a large heart at the time of a bout of pneumonia in January 1957. The patient had had no episodes suggesting failure nor had she been noted to tire on exertion. At the time of this examination she was fairly well developed, weighing 43 1/2 pounds, and in no acute distress. An apical impulse in the anterior axillary line in the fifth intercostal space suggested cardiac enlargement. No thrills could be felt. There was a moderately loud, continuous murmur heard all over the right chest. This murmur was heard to the left of the sternum in the fourth and fifth intercostal spaces and radiated toward the apex. The electrocardiogram revealed evidence of left ventricular hypertrophy. The chest x-ray revealed a large heart with a contour suggesting left ventricular hypertrophy. The pulmonary vascularity was within normal limits. At this time the impression was a ventricular septal defect with the possibility of arteriovenous fistula in the right lung field or a stenosis of a peripheral pulmonary artery on the right. The child's blood pressure was 110/58 in the right arm and 150/54 in the leg.

The child was then referred to the Children's Hospital for cardiac catheterization in February 1960 (table 1). The pulmonary blood flow to systemic blood flow ratio was 2.8/1 with a left-to-right shunt at the atrial level. The pulmonary artery pressure was measured at 55/15 mm. Hg with a mean of 28 mm. Hg. No evidence of peripheral stenosis in the pulmonary artery could be demonstrated (fig. 1).

Because of the unexplained clinical finding of a continuous murmur throughout the right chest, a selective angiocardiogram was performed. This demonstrated nicely the aneurysmal dilatation of the right coronary artery with flow into the right atrium (fig. 2). Following this procedure, plans were made to re-admit the child to the Children's Hospital for cardiac surgery.

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Diagram of catheterization data. Circles show percent oxygen saturation.

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Selective angiocardiogram showing the communication between base of aorta and coronary sinus. The aneurysmal dilatation of the fistula is seen as a round opaque mass on the right of the aorta in the anteroposterior view (top) and posterior to the aorta in the lateral view (bottom).

Table 1
Right Heart Catheterization

<table>
<thead>
<tr>
<th>Location</th>
<th>Per cent O₂ saturation</th>
<th>Pressure (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava (X3)</td>
<td>51-58</td>
<td>..</td>
</tr>
<tr>
<td>Right atrium (High)</td>
<td>76</td>
<td>7</td>
</tr>
<tr>
<td>Right atrium (Low)</td>
<td>84-85</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>73-74</td>
<td>54/0</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>74-78</td>
<td>55/15 (28)</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>97</td>
<td>145/80 (100)</td>
</tr>
<tr>
<td>Pulmonary capillary</td>
<td>..</td>
<td>13</td>
</tr>
</tbody>
</table>

Pulmonary blood flow—3.94 L./min.
Systemic blood flow—1.41 L./min.
Left-to-right shunt—2.53 L./min.

Operation

On July 7, 1961, the patient was anesthetized with endotracheal cyclopropane and a median sternotomy incision made. A striking, continuous thrill was present over the apex of the right atrium before the pericardium was opened. This was found to be sharply localized between the right atrial appendage and the base of the aorta. The right atrium was two times normal size. The first 2 to 3 cm. of the right coronary artery were markedly enlarged, but the distal branches were not unusual (fig. 3). Through the posterior wall of the right atrium, the tense coronary sinus could be felt under greatly increased pressure. Localized point pressure over the origin of the right coronary artery completely stopped the thrill.

When it became apparent that the fistula did not terminate in the lumen of the right atrium proper, the preliminary preparations for extracorporeal circulation were abandoned. Dissection was begun in the aorto-atrial groove overlying the origin of the right coronary artery. The fistulous communication was soon isolated and measured 1 cm. in length. It originated 0.5 cm from the coronary ostium and terminated directly into a greatly dilated tributary of the coronary sinus (fig. 4). Three suture ligatures of 4-0 arterial silk were used to obliterate the fistula. These were placed as purse string sutures at each end with a mattress suture in the middle (fig. 3). The pleural spaces were not entered. The sternotomy was closed with wire without mediastinal drainage. The patient’s postoperative hospital course was unremarkable.

In the postoperative follow-up no cardiac murmurs have been heard. The child is asymptomatic and developing normally.

Discussion

Continuous cardiac murmurs at the base of the heart are most often due to a patent ductus arteriosus. In addition, a long list of other lesions has been given, notably
peripheral pulmonary stenosis, ruptured aneurysm of the sinus of Valsalva, aortic-pulmonary window, and ventricular septal defect with aortic insufficiency. In those cases in which the murmur is atypically located to the right of the sternum, as noted in this patient, selective angiocardiography or retrograde aortography is usually diagnostic.

**Indications for Operative Correction**

In view of the relative simplicity of the surgery required for coronary artery fistulas and the potential hazard when there are definite indications of physiologic abnormality, as noted in this patient, surgical correction seems justified. In the majority of cases of coronary fistulas previously reported, the presence of the lesion was incidentally discovered at postmortem examination. The apparent benignity of the condition is no contraindication to correction when the defect is discovered in childhood.1 There does not seem to be any indication that a reasonable delay is harmful, but we see no advantage in postponing operative intervention beyond puberty.

Of the 74 fistulas of the coronary arteries previously reported,2 the vast majority originated in the left coronary artery. There are less than a dozen reports of communications between the right coronary artery and the right atrium and the majority are autopsy reports.3-11 The first report of a fistula between the right coronary artery and the coronary sinus is a necropsy description by Halpert in 1930.7 This was an incidental finding in a 54-year-old man who died of metastatic carcinoma of the stomach. The only other similar case that we have found was described by Sanger in 1959.10 The exact entry of the right coronary artery fistula into the right atrium was not determined in this case but ligation of the aneurysmal mass of vessels obliterated the shunt and to date the child has done well. As in other types of vascular fistulas, surgical repair of the right coronary artery-coronary sinus communication in this patient presented no special technical problem. The right coronary artery was not ligated and should have normal flow. The coronary sinus was also left intact.

**Summary**

Congenital coronary arteriovenous fistula is easily confused with patent ductus arterio-
sus. An illustrative case is presented which emphasizes the special tests that are necessary to establish the diagnosis. Finally, the successful surgical correction of this abnormality is described.

Acknowledgment
The authors wish to acknowledge the significant contributions of Dr. Leonard Leight (Department of Medicine) and Dr. Lawrence A. Davis (Department of Radiology) in the diagnosis and successful treatment of this child.

References

The "Commentarii" of William Heberden
Very few of Heberden's manuscripts have survived.... The original Latin manuscript of the "Commentarii" is preserved in the Royal College of Physicians. This library also has the manuscript of his Goulstonian Lecture of 1749.... Some of his letters are preserved in the British Museum and the Royal College of Surgeons. To this short list may be added the three volumes of manuscripts found in South London in 1927.

The material in these three volumes throws no new light on the details of his life, but confirms the indications given in the "Commentarii" of his methods of work, and fills in the details of a broad picture of a man intensely interested in the practical side of medicine. The moral essays and the translations found in one of these volumes are a revelation of his interests and mode of thought outside of medical channels. The volume on strictly medical subjects, which must antedate by many years his final opinions as expressed in the "Commentarii," shows his power of observation, assimilation and constructive thought in matters medical.—Prefatory Essay, by LEROY CRUMMER. WILLIAM HEBERDEN. An Introduction to the Study of Physic. New York, Paul B. Hoeber, Inc., 1929, p. 33.
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