Congenital Communication of a Coronary Artery with a Cardiac Chamber or the Pulmonary Trunk ("Coronary Artery Fistula")

By Henry N. Neufeld, M.D., Richard G. Lester, M.D., Paul Adams, Jr., M.D., Ray C. Anderson, M.D., C. Walton Lillehei, M.D., and Jesse E. Edwards, M.D.

The communication of a coronary artery with a cardiac chamber or the pulmonary trunk, although an uncommon anomaly, is important for several reasons. It enters into the differential diagnosis of those conditions associated with a continuous precordial murmur, among them patent ductus arteriosus, aorticopulmonary septal defect, ventricular septal defect with aortic insufficiency, supravalvar pulmonary arterial stenosis, ruptured aneurysm of an aortic sinus, and congenital absence of the pulmonary valve. Moreover, the anomaly may become complicated by bacterial infection or congestive cardiac failure or both. Ordinarily, it is amenable to cure by interruption of the fistulous tract.

In this report we describe six cases observed at the University of Minnesota Hospitals and compare them with those reported in the literature.

The six patients, three male and three female, ranged in age from 17 months to 53 years (table 1).

Of the six patients, five underwent surgical treatment; all five are living. The sixth patient (case IV) did not receive surgical therapy; the diagnosis was confirmed at necropsy.

Pathologic Features

There are two types of gross communication between a coronary artery and a cardiac chamber. The first type is characterized by an intact ventricular septum (or a very small ventricular septal defect) and atresia either of the pulmonary or the aortic valve. Under these circumstances, and in the presence of a competent corresponding atrioventricular valve, enlarged sinusoids of the ventricle proximal to the atretic semilunar valve penetrate the myocardium. These sinusoids converge in or near the epicardium to form a single vessel leading from the obstructed ventricle. In the epicardium, this anomalous vessel joins with the usual branches of the coronary arteries. Then, blood from the obstructed ventricle is forced into the coronary arteries, which receive this blood in addition to their supply from the aorta. When pulmonary atresia is present, venous blood is therefore delivered to the coronary arteries, while when aortic atresia is present, blood fully saturated with oxygen is delivered from the left ventricle directly into the coronary arteries by way of the channels mentioned.

In all the six cases here reported the anomalous coronary arterial communication was of the second type. In it a coronary artery communicates with a cardiac chamber or pulmonary arterial trunk without other cardiac malformation, except coincidentally. In our six cases the abnormal coronary artery was the right in two cases and the left in four cases. The sites of termination of the coronary artery were as follows: (1) when the right artery was involved—right ventricle, one case (case I) and right atrium, one case (case VI); (2) when the left artery was involved—right atrium, one case (case IV), right ventricular cavity, one case (case II),...
Table 1

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Respiratory infections</th>
<th>Dyspnea</th>
<th>Congestive heart failure</th>
<th>Systolic thrill</th>
<th>Continuous murmur</th>
<th>Second sound pulmonary area</th>
<th>Systemic blood pressure</th>
<th>Electrocardiogram</th>
<th>Coronary artery</th>
<th>Site of termination</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>10</td>
<td>F</td>
<td>+++</td>
<td>+*</td>
<td>–†</td>
<td>+</td>
<td>Gr. III 4th LICS</td>
<td>Normal</td>
<td>100/60</td>
<td>–170</td>
<td>R.V. Diast. Overload</td>
<td>Right</td>
</tr>
<tr>
<td>II</td>
<td>1 yr. 5 mo.</td>
<td>M</td>
<td>++</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>Gr. III at apex area</td>
<td>++</td>
<td>140/80</td>
<td>+ 90*</td>
<td>None</td>
<td>Left</td>
</tr>
<tr>
<td>III</td>
<td>53</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>–</td>
<td>Gr. III 2-3rd LICS</td>
<td>Normal</td>
<td>160/100</td>
<td>+ 60*</td>
<td>Left Vent.</td>
<td>Left</td>
</tr>
<tr>
<td>IV</td>
<td>43</td>
<td>M</td>
<td>–</td>
<td>–</td>
<td>++</td>
<td>–</td>
<td>4th LICS</td>
<td>+</td>
<td>160/75</td>
<td>+ 30*</td>
<td>Left Vent.</td>
<td>Left</td>
</tr>
<tr>
<td>V</td>
<td>19</td>
<td>M</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>3rd LICS</td>
<td>+</td>
<td>125/85</td>
<td>+ 90*</td>
<td>–</td>
<td>Left</td>
</tr>
<tr>
<td>VI</td>
<td>19</td>
<td>F</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>+</td>
<td>2 RICS</td>
<td>+</td>
<td>130/60</td>
<td>+ 90*</td>
<td>–</td>
<td>Right</td>
</tr>
</tbody>
</table>

RA, right atrium; RV, right ventricle; PT, pulmonary arterial trunk; LICS, left intercostal space; RICS, right intercostal space.

*++, Condition present. †–, Condition not present.

In early prenatal life, the coronary arteries communicate with the veins through a classic capillary network, they also give off branches to the intertrabecular spaces that communicate with the arteriovenous communication.

**Embryology**

In our series, in one of our patients (case V), a segment of the coronary artery was removed for histologic examination. The segment revealed marked thickening of the media with prominent muscle bundles interspersed with numerous elastic fibers. Nonspecific focal fibrous thickening was also present (fig. 1). These features are common in arteries in any site that lies proximal to an arteriovenous communication. A single opening is often described at the site of an anomalous communication (table 2). Although an attempt at anamnestic information was made to review all reported cases, some may have been overlooked. In total, 50 cases have been reported, although anomalous termination of coronary arteries and pulmonary trunk, two cases (cases III and IV), and anomalous termination of embryologic sites of coronary arteries (table 2).
cate with the ventricular cavities. In later prenatal life, the intertrabecular spaces shrink to form the sinusoids, which represent communications between the veins and coronary arteries, on the one hand, and the cardiac chambers on the other.\textsuperscript{43} Abnormally large connections of the type here considered appear to represent persistence of the large intertrabecular spaces, which connect with a ventricular cavity, on the one hand, and with a coronary artery, on the other.

A different explanation for anomalous communication between a coronary artery and the pulmonary trunk exists. This anomaly apparently occurs when one or several coronary arteries arise from the pulmonary trunk. The accessory artery or arteries in turn make communication with branches of one or both normally arising coronary arteries. As flow from the high pressure (aortic) area is carried into the low pressure (pulmonary) area, the communication enlarges.

**Clinical Features**

The natural history of this malformation is not well known. In most cases reported in the literature the patients reached adult life. Only occasionally did death occur in infancy or childhood.

Symptoms seem to depend upon the magnitude of the shunt. Among our patients, three gave histories of recurrent respiratory infections. Congestive cardiac failure was observed in two patients (cases III and IV) and was the cause of death in one of them (case IV). In another patient (case III), signs of congestive cardiac failure disappeared after surgical interruption of the anomalous channel.

Some patients may manifest symptoms and signs of coronary insufficiency. In case IV, the patient was a 43-year-old man whose chief complaint was angina. Obviously, in this age group it is difficult to determine whether angina is related to the malformation itself or to concomitant coronary atherosclerosis. In other cases, bacterial infection may develop in the abnormal communication.\textsuperscript{47} Steinberg and associates\textsuperscript{44} reviewed 22 reported cases of coronary arteriovenous fistula and added one case diagnosed by angiocardiography. In only two cases was death directly attributed to the fistula; in the others it was due to nonrelated causes.

In those cases in which the anomalous termination is into the left ventricle, only a diastolic murmur is anticipated. In cases involving the other possible sites of termination, a continuous murmur frequently occurs; in fact, it is the most consistent abnormal physical sign among reported cases, and was noted in all six cases reported here.

The location of maximal intensity of the continuous murmur seems to depend upon the site of the abnormal communication, i.e., its position usually corresponds with the anatomic position of the chamber or vessel participating in the anomalous connection. We observed the association between a continuous murmur and a systolic thrill only in two patients (cases I and VI), but this association is commonly reported in the literature. The second sound at the pulmonic area was accentuated in all six of our patients and was particularly prominent in four (cases II, IV, V, and VI). The absence of a murmur in Scott's case\textsuperscript{44} is perhaps explained by the observation at necropsy that a thrombus had occluded the fistula.

\textbf{Figure 1}

\textit{Case I. Photomicrograph of right coronary artery proximal to its entrance into the right ventricle. Hypertrophy of the media. Numerous elastic fibers course between bundles of muscle. The intima (above) shows mild nonspecific fibrous thickening. Elastic-tissue stain.}
Gasul and associates documented with phonocardiography their observation that the diastolic component of the continuous murmur was louder than the systolic component in patients with fistulas that communicated with the right ventricle. This diastolic accentuation contrasts with the systolic accentuation usually observed in patent ductus arteriosus.

Electrocardiographic Features

In anomalous communication between a coronary artery and a cardiac chamber or the pulmonary trunk, the electrocardiogram may show various abnormalities, but no specific diagnostic patterns are present.

Electrocardiograms were obtained for each of our six patients. In two cases (cases III and IV), a pattern of left ventricular overload was observed. One case (case I) showed right ventricular diastolic overload, and one case (case VI) showed flattening of the T waves in all leads. In the remaining two cases no electrocardiographic abnormalities were noted.

The electrocardiographic findings seem to depend upon the anatomic situation and associated alterations in hemodynamics. If the left-to-right shunt is small and insignificant, no specific abnormal findings will appear in the electrocardiogram. If, however, a wide anomalous communication and a large shunt exist, signs of left ventricular hypertrophy may be found. In case IV of our group, the left-to-right shunt was shown to be more than 8 liters per minute per square meter of body surface and the electrocardiogram revealed signs of left ventricular overwork. In cases in which the coronary artery is anomalously connected with the right atrium, both left ventricular hypertrophy and right ventricular volume overload pattern may occur, as in the case reported by Edwards and associates.

Quite apart from shunts potentially affecting the electrocardiogram, the possibility of alterations resulting from associated myocardial ischemia should be considered. Usually no such alterations are present, however. In case IV of our series, although the patient presented a history of anginal pain, electrocardiogram did not reveal changes in the ST-T segments.

In one case reported by Colbeck and Shaw, involving anomalous communication between the right coronary artery and right atrium in an 80-year-old man, the electrocardiogram showed atrial fibrillation and left bundle-branch block. At necropsy the heart weighed 554 Gm., and left ventricular hypertrophy

Table 2

<table>
<thead>
<tr>
<th>Coronary Artery of Origin and Site of Anomalous Termination; 50 Cases Reported in this Study and in the Literature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coronary involved</td>
</tr>
<tr>
<td>-------------------</td>
</tr>
<tr>
<td>Right</td>
</tr>
<tr>
<td>Right</td>
</tr>
<tr>
<td>Right</td>
</tr>
<tr>
<td>Total right</td>
</tr>
<tr>
<td>Left</td>
</tr>
<tr>
<td>Left</td>
</tr>
<tr>
<td>Left</td>
</tr>
<tr>
<td>Left</td>
</tr>
<tr>
<td>Left</td>
</tr>
<tr>
<td>Total left</td>
</tr>
<tr>
<td>Total left and right</td>
</tr>
</tbody>
</table>

*Roman numerals indicate case number of cases in this communication.
†This case was reported previously.

Circulation, Volume XXIV, August 1961
and dilatation were noted. The important functional disturbance of this condition is the run-off of blood from the coronary arterial system similar to that in other arteries that lie proximal to arteriovenous or arteriovenous-like fistulas. Consequently, when a coronary artery communicates with a right-sided cardiac chamber, cardiac catheterization may reveal high oxygen saturation of the blood in that chamber receiving blood from the coronary fistula. When the abnormal vessel communicates with the left side of the heart, no abnormality in oxygen saturation occurs.

Hemodynamic studies were performed in four of our six cases. A summary of the observations is shown in table 3. In each case an increase in oxygen saturation was detected in the chamber or vessel receiving the abnormal communication (right atrium, one case; right ventricle, two cases; pulmonary trunk, one case). Calculated shunts varied from a minimum of 0.6 to 8.7 liters per minute per square meter of body surface. A similarly high value for a left-to-right shunt was obtained in one of Gasul’s cases.\textsuperscript{16} Pulmonary arterial and right-sided intracardiac pressures were within normal limits in two cases.

In the anomaly under discussion, diversion of aortic blood into the coronary system is greater than normal. In the normal cases about 10 per cent of aortic blood flows into the coronary arterial system, but in patients with this malformation more than half the aortic blood may be diverted into the abnormal coronary artery. Most of this blood is not directed into the myocardial capillaries, which constitute a zone of high resistance; instead, the major part of the blood is directed through the low-resistance channel, the anomalous communication. In addition, blood from the other coronary artery may be diverted through intercoronary anastomoses to the fistula. Thus, for example, in a case in which the right coronary artery connects anomalously with a cardiac chamber, the myocardium in the distribution of the left coronary artery may be ischemic from the diversion of blood toward the fistula.

Hemodynamic studies may readily permit the identification of the site of anomalous communication of a coronary artery with a right-sided chamber or the pulmonary trunk. They do not, however, reveal the anatomic basis for the shunt. Ruptured aneurysm of an aortic sinus, ventricular septal defect with aortic insufficiency, and aorticopulmonary communications—all these may yield findings similar to those observed in anomalous communication of a coronary artery with a right-sided cardiac chamber or the pulmonary trunk. Precise anatomic diagnosis requires selective aortography.

In Mozen’s case,\textsuperscript{57} in which the fistual communicated with the left atrium, the findings of catheterization of the right side of the heart were normal.

**Roentgenographic Features**

Roentgenographic examination of the thorax revealed moderate to severe cardiomegaly.
in four cases (cases II, III, IV, and V, fig. 2). In the two remaining patients (cases I and VI), the size of the cardiac shadow was within normal limits. The pulmonary arterial segment was prominent in all cases, and the peripheral pulmonary arterial vasculature was moderately to markedly increased in all but one patient (case VI).

The aortic knob was prominent in all cases. From conventional films, one cannot distinguish the findings in this anomaly from those in other conditions in which left-to-right shunts and continuous machinery murmurs occur. In general, the roentgenographic appearance seems to depend on the hemodynamic alterations of the particular case.

Gasul and associates,16 summarized the roentgenographic findings in 28 cases, including five of their own. In nine cases these were considered to be normal; in the other 19 cases, the common findings were enlargement of the left ventricle, dilatation of the pulmonary arterial segment, and prominence of the pulmonary vasculature.

Although retrograde aortography by means of an injection through a radial artery is a time-honored procedure for diagnosing patent ductus arteriosus, it is very often unsatisfactory for diagnosing lesions originating in the ascending aorta; these lesions include: anomalous communication of a coronary artery with a cardiac chamber or the pulmonary trunk, aortieopulmonary septal defect, persistent truncus arteriosus, or ruptured aneurysm of an aortic sinus of Valsalva.

In most cases, opaque material reaching the arch of the aorta by way of a retrograde injection follows the flow of blood in the aorta distally into the descending aorta. Thus the entire ascending aorta, including its sinuses, is usually not satisfactorily visualized by this method. This phenomenon was well demonstrated in one of our patients when two retrograde aortograms failed to demonstrate the anomalous termination of a coronary artery.

We prefer to use selective aortography, a procedure in which radiopaque material is injected into the base of the aorta through a catheter. Selective aortography was performed in three cases (cases I, II, and III): In case I, the aorta was shown to be dilated and densely opacified (fig. 3). The right coronary artery, which was unusually dilated, measured approximately 1½ to 2 cm. in diameter. Multiple saccular aneurysms were seen along the course of the anomalous vessel. Sequential filling of the right ventricle and pulmonary artery was observed following demonstration of anomalous communication.
of the right coronary artery with the right ventricle.

In case II (fig. 4), a large, tortuous, irregularly dilated vessel arising above the left aortic sinus became apparent immediately after filling of the aortic sinuses. This channel progressed to the left and passed downward in the region of the anterior interventricular groove. Filling of this channel was followed by opacification of the right ventricular chamber and then of the pulmonary artery in normal sequence.

In case III, opacification of the left coronary artery was followed by filling of the main pulmonary artery, without opacification of the right ventricle.

Summary

Anatomic, clinical, hemodynamic, and roentgenographic findings in six patients with congenital communication of a coronary artery with a cardiac chamber or the pulmonary trunk are presented. The literature is reviewed.

A coronary artery may communicate anomalously with any of the cardiac chambers, more commonly with those on the right side. In the six cases presented, the right coronary artery communicated with the right atrium in one case and with the right ventricle in another. The left coronary artery communicated with the right atrium and right ventricle in one case each, and with the pulmonary trunk in two cases.

The most striking feature observed clinically was a continuous murmur. If a continuous murmur is localized in an area atypical for patent ductus arteriosus, the diagnosis should be suspected. Conventional roentgenographic and electrocardiographic studies yielded no specific diagnostic features. The results of cardiac catheterization may reveal a left-to-right shunt, but they are diagnostically useful only when correlated with clinical findings.

The only precise method of demonstrating the abnormality is by means of selective aortography performed by injecting medium into the very origin of the aorta.

Cure is possible by surgical interruption of the fistulous tract.
References


6. Johnson, J.: Quoted by Davis and associates.2


14. Engle, M. A.: Quoted by Gasul and associates.3


23. 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 cirrhotic dilatation of the vessels. J. Anat. & Physiol. 20: 26, 1886.


"CORONARY ARTERY FISTULA"


On Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves

By Dominic John Corrigan, M.D.

One of the Physicians to the Charitable Infirmary, Jervis Street, Dublin; Lecturer on the Theory and Practice of Medicine; Consulting Physician to St. Patrick's College, Maynooth.

... From its singular and striking appearance, the name of visible pulsation is given to this beating of the arteries. It is accompanied with bruit de soufflet in the ascending aorta, carotids, and subclavians; and in the carotids and subclavians, where they can be examined by the finger, there is felt fremissement, or the peculiar rushing thrill, accompanying with bruit de soufflet each diastole of these vessels. These three signs are so intimately connected with the pathological causes of the disease, and arise so directly from the mechanical inadequacy of the valves, that they afford unerring indications of the nature of the disease.
Congenital Communication of a Coronary Artery with a Cardiac Chamber or the Pulmonary Trunk ("Coronary Artery Fistula")
HENRY N. NEUFELD, RICHARD G. LESTER, PAUL ADAMS, JR., RAY C. ANDERSON, C. WALTON LILLEHEI and JESSE E. EDWARDS

Circulation. 1961;24:171-179
doi: 10.1161/01.CIR.24.2.171
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1961 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/24/2/171

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/