Review of Single Coronary Artery with Report of 2 Cases

By JOHN CHANDLER SMITH, M.D.

The paper includes a discussion of all published cases of single coronary artery and a consideration of the embryologic development and physiologic significance of this anomaly. The incidence and types of associated cardiac diseases are reviewed. Two additional case reports of this anomaly are included.

The occurrence of a single coronary artery is such a rare cardiac anomaly that presentation of 2 additional cases is of interest. The first case is unique in that the patient was the oldest in whom such an anomaly has been reported. In addition, pertinent data on 43 previously reported cases are presented.

Case Reports

Case 1.—The patient was a white woman, 80 years of age, who entered University Hospitals of Cleveland with complaints of constipation and severe nausea for the past twenty-four hours and sharp attacks of pain over the lower abdomen of twelve hours' duration. She had never experienced chest pain, shortness of breath or edema of the ankles. Physical examination revealed a firm mass in the rectum and a sigmoid colostomy was performed on the day of hospital admission. On the thirteenth hospital day an abdominoperineal resection was performed and examination of the specimen disclosed a partially differentiated adenocarcinoma of the rectum. Metastases were not found. Edema of the legs and ankles developed on the fourth hospital day. The patient became comatose and died on the eleventh hospital day.

Autopsy (No. 15031): The heart weighed 340 grams. The epicardium was smooth and glistening. The muscle of the ventricles was uniformly brownish red and moderately firm throughout. The endocardium was smooth and glistening except over the posterior wall of the left atrium where there was a poorly demarcated focus of fine wrinkling. The leaflets of the mitral valve were slightly thickened and opaque along the free border. The cusps of the aortic valve were smooth and pliable and revealed moderate subcommissural adhesions.

A saccular dilatation, 0.6 cm. in diameter, was present in the wall of the right aortic sinus at the normal site of origin of the right coronary artery. A single coronary artery originated from the left aortic sinus and divided into an anterior descending and a circumflex branch. The former divided once and both branches extended obliquely over the anterior surface of the left ventricle, the larger continuing to the apex where it could be followed over the posterior surface of the left ventricle for 2 cm. Cross sections of the first portion of the larger anterior descending branch revealed yellowish gray intimal plaques that nearly occluded the lumen. The circumflex branch of the left coronary artery extended around the posterior surface of the left ventricle along the atrioventricular groove to the acute margin of the heart and continued to the anterior surface of the right ventricle to the base of the right auricular appendage. The course of this vessel measured 25 cm. from aortic orifice to the base of the right auricular appendage (fig. 1). Cross sections revealed a patent lumen throughout with slight focal thickening of the intima by yellowish gray plaques. Small branches extended over the posterior surface of both ventricles and over the anterior surface of the right ventricle.

Histologic examination of the myocardium revealed moderately large muscle fibers with nuclei that were slightly enlarged and occasionally rectangular. The myocardium was not otherwise remarkable. Histologic sections of the first portion of the larger branch of the left anterior descending coronary artery revealed marked thickening of the intima and subintimal deposition of a large amount of eosinophilic material containing acicular spaces. The lumen was reduced to a slit.

The pathologic diagnoses included recent abdominoperineal resection for partially differentiated adenocarcinoma of the rectum, acute fibrino-purulent peritonitis, recent sigmoid colostomy and absence of the anterior abdominal wall. There was a single left coronary artery. In addition, there were healed nondeforming endocarditis of the mitral and aortic valves, bronchopneumonia of the right and left lungs, chronic cholecystitis with cholelithiasis and slight arteriolar nephrosclerosis.

Case 2.—The patient was a white woman 66 years of age who entered University Hospitals of Cleveland with complaints of lethargy, slurring of speech and weakness of the left arm of two days' duration. She had been diabetic for the past eight years and had been treated with 15 units of insulin daily. There had been no chest pain, ankle edema or shortness of breath.

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FIG. 1.—The distribution of the single left coronary artery of Case 1.

FIG. 2.—The distribution of the single left coronary artery of Case 2 with an anomalous branch extending to the right ventricle.

Urinalysis revealed 4 plus sugar, 4 plus acetone and 3 plus di-acetic acid. The blood sugar was 224 mg. per 100 cc. and the carbon dioxide combining power of the plasma was 58 volumes per cent.
Intravenous infusion of 5 per cent glucose in normal saline with 35 units of regular insulin was given. Small doses of insulin were given throughout the night and by the second hospital day the urine was free of acetone. However, the temperature gradually rose to 40 C. and the patient died on the third hospital day.

Autopsy (No. 8022): The heart weighed 250 grams. The epicardium was smooth and glistening. The myocardium was moderately firm and light brownish red throughout. The free margins of the tricuspid and mitral valves were thickened and opaque. There were slight subcommissural adhesions between the cusps of the aortic valve. The endocardium was smooth and glistening except over the posterior wall of the left atrium where there was a focus of fine wrinkling and gray opacity.

A single coronary artery originated from the left aortic sinus and divided into an anterior descending branch and a circumflex branch. Approximately 1 cm. from the origin of this single coronary artery, a moderately large branch extended to the right, passing anterior to the pulmonary conus to be distributed over the anterior and lateral surfaces of the right ventricle (fig. 2). The circumflex branch extended around the atrio-ventricular fold to send a branch down the obtuse margin of the left ventricle and a branch down the posterior surface of the interventricular septum. From the latter, small branches extended to the posterior surface of the right ventricle. Cross sections revealed yellow intramural deposits that encroached moderately on the lumens of all of these arteries.

Microscopic examination of the myocardium revealed oval nuclei within the muscle fibers. Foci of fibrosis were not seen. Histologic sections of the coronary arteries revealed needle-shaped spaces within subintimal depositions of eosinophilic material that encroached moderately upon the lumens. There was a large amount of glycogen within the epithelial cells of the renal tubules. There was moderate fat infiltration and marked atrophy and interstitial fibrosis of the pancreas.

The pathologic diagnoses included moderate arteriosclerosis of a single left coronary artery, bronchopneumonia of the right and left lungs, pronounced fibrosis and atrophy of the pancreas, moderate arteriolar nephrosclerosis and acute hemorrhagic cystitis.

**DISCUSSION**

According to Banchi, a case of single coronary artery was reported by Thebesius in 1716. In 1827, Mayer described, as an incidental anomaly in a fetus of 7 months' gestation with cor bilocular, a single coronary artery that arose from the right carotid artery and extended to the heart to divide into two branches to supply the myocardium. Since 1716, 43 cases have been reported. With the 2 cases here described, a total of 45 cases of single coronary artery is now on record.

The definition of a heart with a single coronary artery is one in which the entire myocardium is nourished by an artery, regardless of distribution, that arises by one ostium from an arterial trunk. In all but 2 of the cases thus far reported, the single coronary artery has arisen from the aorta. The case of Mayer is mentioned above. In the case of Tow, examination of a white female infant, 5 months old, revealed a cor bilocular with a single vessel arising from the pulmonary artery that divided into two branches to supply the myocardium. No arteries issued from the ascending aorta. In the case of Forester, examination of a newborn white male infant revealed no arteries originating from the sinuses of Valsalva but a single coronary artery arising from the inferior aspect of the arch of the aorta. In all other reported cases the single vessel has arisen from the aorta within one of the sinuses of Valsalva.

According to the definition of single coronary artery here set down, the following cases, although included in other reviews, have been omitted here. The case of Grätzer and the case of Sanes are excluded because, in each, three separate coronary arteries issued from a single aortic sinus. The case of Bland, White and Garland is also excluded because 2 closely adjacent coronary arteries arose from the same aortic sinus. The 21 cases reviewed by Soloff are not included because in each the heart was supplied by two coronary arteries although one or both arose from the pulmonary artery.

With these deletions, the literature embodies 43 acceptable cases of single coronary artery. Because of embryologic considerations relative to the formation of this anomaly, these cases are conveniently classified into three types according to the distribution of the single coronary artery. The first type includes those cases in which the single vessel follows the course of only the normal right or left coronary artery. This type is illustrated in figure 1 and the 10 reported cases of this type are assembled in table 1. The second type of single coronary
artery includes those cases in which the single vessel arises by one ostium but divides so that branches are present in the distribution of both the right and left coronary arteries. This distribution is illustrated in figure 2 and the 17 cases of this type are assembled according to patient age in table 2. In 1938, Krumbhaar and Ehrich\(^2\) included as a group those cases of single coronary artery in which the distribution of the vessel was so atypical that it could not be compared in this regard with either the right or the left coronary artery. The 12 cases of this type are assembled in table 3. Also included in table 3 are 3 cases in which myocardium. Both arteries pass at first to the bulbus cordis and then spread out over the heart, uniting with the capillary network and intertrabecular spaces in the developing myocardium. According to Grant,\(^3\) the same description of development applies to the human heart.

Most authors agree that the majority of cases of single coronary artery are caused by one of two developmental anomalies. The first is a congenital absence of one coronary artery anlage. In cases presumably of this type, assembled in table 1, the single vessel follows the normal course of the artery it represents.

### TABLE 1.—Single Artery Present in the Distribution of Only One Coronary Artery (Fig. 1)

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Single Artery Present</th>
<th>Weight of Heart</th>
<th>Autopsy</th>
<th>Year</th>
<th>Author</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>4 days</td>
<td>M</td>
<td>L</td>
<td>30</td>
<td>Truncus solitarius</td>
<td>1930</td>
<td>Shapiro(^4)</td>
</tr>
<tr>
<td>2.</td>
<td>33 years</td>
<td>M</td>
<td>L</td>
<td>520</td>
<td>Cerebral hemorrhage</td>
<td>1930</td>
<td>Petren(^5)</td>
</tr>
<tr>
<td>3.</td>
<td>35 years</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>Dissecting aortic aneurysm‡</td>
<td>1947</td>
<td>Roberts and Louba(^6) (case 7)</td>
</tr>
<tr>
<td>4.</td>
<td>37 years</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>Subacute bacterial endocarditis‡</td>
<td>1922</td>
<td>Plaut(^7)</td>
</tr>
<tr>
<td>5.</td>
<td>39 years</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>Pneumonia</td>
<td>1935</td>
<td>Kochel(^8)</td>
</tr>
<tr>
<td>6.</td>
<td>42 years</td>
<td>F</td>
<td>L</td>
<td>430</td>
<td>Rheumatic myocarditis</td>
<td>1940</td>
<td>Madox and Ibister(^9)</td>
</tr>
<tr>
<td>7.</td>
<td>44 years</td>
<td>F</td>
<td>L</td>
<td>400</td>
<td>Pulmonary embolus‡</td>
<td>1938</td>
<td>Krumbhaar and Ehrich(^10) (case 1)</td>
</tr>
<tr>
<td>8.</td>
<td>45 years</td>
<td>M</td>
<td>R</td>
<td>480</td>
<td>Lobar pneumonia</td>
<td>1940</td>
<td>King(^11)</td>
</tr>
<tr>
<td>9.</td>
<td>63 years</td>
<td>M</td>
<td>L</td>
<td>150</td>
<td>Carcinoma of stomach</td>
<td>1937</td>
<td>Riehler(^12) (case 1)</td>
</tr>
<tr>
<td>10.</td>
<td>66 years</td>
<td>F</td>
<td>L</td>
<td>340</td>
<td>Carcinoma of colon</td>
<td>1947</td>
<td>Geever and Ravin(^13)</td>
</tr>
</tbody>
</table>

* Left or right side.

† In addition to the diagnoses listed, there was a saucellar dilatation of the aortic sinus from which the missing coronary artery normally arises.

In 1882, Hyrtl\(^14\) stated that cases of absent coronary artery were limited to examples of this type. It was his opinion that cases in which one artery supplied the heart in the distribution of both normal right and left coronary arteries were examples of misplacement rather than absence of one coronary artery anlage. Other authors,\(^27, 31, 32\) however, suggested that in the absence of one coronary artery, the remaining vessel might develop compensatory branches that would follow the course of the missing vessel and act as functional substitutes. As both of these suggestions are tenable, and neither has been disproved, the distribution of the single vessel cannot
be used as a criterion to distinguish absence from misplacement of one coronary artery anlage.

The second developmental defect accepted as a cause of single coronary artery is misplacement of one coronary artery anlage so that this anlage fuses with the first portion of the remaining normal vessel. This has been attributed to misplacement of the septum dividing the truncus communis into pulmonary artery and aorta with crowding together of the coronary artery anlagen. All authors recognize that cases of single coronary artery are examples of misplaced coronary anlagen and that the essential lesion is not an absence of the vascular supply. Krumbhaar and Ehrich had no way of deciding whether the anlage was absent or misplaced but stated that the small saccular dilatation of the aortic sinus at the site from which the missing vessel normally arises suggests that this represents a remnant of an absent coronary anlage and that the entire coronary arterial system in these cases arises from one coronary anlage. However, of the 8 cases in which such a saccular dilatation was present, the distribution of the single vessel in 2 cases was that of both the

remaining normal vessel. This has been attributed to misplacement of the septum dividing the truncus communis into pulmonary artery and aorta with crowding together of the coronary artery anlagen. The distribution of the single vessel in these cases is that of the normal right and left coronary arteries. These cases make up table 2. Sanes stated that
normal right and left coronary arteries. Since the question of absence or misplacement of one coronary anlage may be considered unsettled, it is preferable at present to record cases of single coronary artery as such, rather than as cases of anomalous origin of one coronary artery or as cases of absent coronary artery.

There were 27 cases of single coronary artery occurring in adults. Of these, 18 were men and 8 were women. In one case the sex of the patient was not stated. The average age of the adult cases was 45 years and the oldest patient was 80. There were 13 cases of single right coronary artery and 15 cases of single left coronary artery. In 3 cases occurring in adults, the single artery was not named or described. In 7 cases in which the single vessel was present in the distribution of only one normal coronary artery, the average heart weight was 380 grams, with extremes of 150 and 520 grams, respectively. Similar data in the 7 cases in which the single vessel was present in the distribution of both coronary arteries revealed that the average heart weight was 505 grams, with extremes of 240 and 930 grams, respectively. Signs or symptoms of decreased cardiac function were not described in any case of single coronary artery in which autopsy examination revealed an otherwise normal cardiovascular system. In none of the cases

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Sex</th>
<th>Single Artery Present</th>
<th>Weight of Heart</th>
<th>Autopsy</th>
<th>Year</th>
<th>Author</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Fetus, 6 months</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>Cor biloculare</td>
<td>1827</td>
<td>Mayer†</td>
</tr>
<tr>
<td>2</td>
<td>Newborn</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>Cor biloculare</td>
<td>1847</td>
<td>Forster‡</td>
</tr>
<tr>
<td>3</td>
<td>Newborn</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>Bicuspid aortic valve</td>
<td>1890</td>
<td>Preiss§ (case 1)</td>
</tr>
<tr>
<td>4</td>
<td>2 days</td>
<td>F</td>
<td>—</td>
<td>—</td>
<td>Cor triloculate biaatriatum</td>
<td>1898</td>
<td>Clark§</td>
</tr>
<tr>
<td>5</td>
<td>3 days</td>
<td>F</td>
<td>R</td>
<td>—</td>
<td>Bronchopneumonia†</td>
<td>1947</td>
<td>Roberts and Loube† (case 8)</td>
</tr>
<tr>
<td>6</td>
<td>4 days</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>Bicuspid aortic valve</td>
<td>1947</td>
<td>Roberts and Loube† (case 9)</td>
</tr>
<tr>
<td>7</td>
<td>7 days</td>
<td>M</td>
<td>R</td>
<td>—</td>
<td>Hypoplasia of left ventricle and aorta†</td>
<td>1924</td>
<td>Henke and Lubarsch‡</td>
</tr>
<tr>
<td>8</td>
<td>3 months</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>—</td>
<td>1931</td>
<td>Geever and Ravin§</td>
</tr>
<tr>
<td>9</td>
<td>3 months</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Bronchopneumonia</td>
<td>1931</td>
<td>Tow§</td>
</tr>
<tr>
<td>10</td>
<td>5 months</td>
<td>F</td>
<td>—</td>
<td>—</td>
<td>Cor biloculare</td>
<td>1931</td>
<td>Kugel†</td>
</tr>
<tr>
<td>11</td>
<td>6 months</td>
<td>F</td>
<td>L</td>
<td>—</td>
<td>Atresia pulmonic artery, absent tricuspid valve</td>
<td>1918</td>
<td>deVries† (case 4)</td>
</tr>
<tr>
<td>12</td>
<td>5 years</td>
<td>M</td>
<td>L</td>
<td>160</td>
<td>Stenosis pulmonic conus, persistent truncus communis</td>
<td>1931</td>
<td>Roberts and Loube† (case 5)</td>
</tr>
<tr>
<td>13</td>
<td>Young adult</td>
<td>M</td>
<td>L</td>
<td>370</td>
<td>Bicuspid aortic valve</td>
<td>1937</td>
<td>Richter‡ (case 2)</td>
</tr>
<tr>
<td>14</td>
<td>22 years</td>
<td>M</td>
<td>L</td>
<td>—</td>
<td>—</td>
<td>1947</td>
<td>Roberts and Loube† (case 3)</td>
</tr>
<tr>
<td>15</td>
<td>38 years</td>
<td>F</td>
<td>L</td>
<td>300</td>
<td>Thrombus of coronary artery, recent infarct of myocardium†</td>
<td>1947</td>
<td>Roberts and Loube† (case 3)</td>
</tr>
</tbody>
</table>

* Left or right side.
† Saccular dilatation of aortic sinus from which missing coronary artery normally arises.

Table 3.—Atypical Distribution of the Single Coronary Artery

Of the 27 adults with single coronary artery the cause of death was reported as related to cardiac disease in 9 cases. Autopsy examinations revealed a recent infarct of the myocardium in 3 cases and an old infarct in one case. The extent of the infarct was described in case 1 of the series of Roberts and Loube.11 Here the right atrium and a large part of the
right ventricle revealed a recent infarct. There was a recent thrombus of the first portion of a single right coronary artery. The diagnoses of the 9 cases in which cardiac disease was present are shown in table 4.

Single coronary artery was reported in 13 infants and one child. In 11 of these cases the distribution of the single vessel was atypical and did not resemble that of either the right or left coronary artery. In 9 of the 14 cases there were other anomalies of the cardiovascular system. These included corn biloculare, bicuspid aortic valve, cor triloculare biaatriatum and hypoplasia of the aorta and left ventricle. Of these 14 cases of single coronary artery, one child lived to the age of 5 years, but of the remainder, the oldest lived to the age of 7 months.

**Table 4.** Cardiac Lesions Found in Cases of Single Coronary Artery

<table>
<thead>
<tr>
<th>Autopsy</th>
<th>Age</th>
<th>Author</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Subacute bacterial endocarditis</td>
<td>37</td>
<td>Plaut</td>
</tr>
<tr>
<td>2. Hemopericardium</td>
<td>37</td>
<td>Roberts and Loube (case 6)</td>
</tr>
<tr>
<td>3. Thrombus of single coronary artery. Recent infarct of myocardium</td>
<td>38</td>
<td>Roberts and Loube (case 3)</td>
</tr>
<tr>
<td>4. Rheumatic myocarditis</td>
<td>42</td>
<td>Maddox and Ibister</td>
</tr>
<tr>
<td>5. Chronic endocarditis of mitral and aortic valves</td>
<td>45</td>
<td>Gallavardin and Ravault</td>
</tr>
<tr>
<td>6. Thrombus of single coronary artery. Recent infarct of myocardium</td>
<td>46</td>
<td>Smith and Graber</td>
</tr>
<tr>
<td>7. Old infarct of myocardium</td>
<td>46</td>
<td>Roberts and Loube (case 4)</td>
</tr>
<tr>
<td>8. Chronic endocarditis of mitral and aortic valves</td>
<td>60</td>
<td>Bochdalek</td>
</tr>
<tr>
<td>9. Thrombus of single coronary artery. Recent infarct of myocardium</td>
<td>62</td>
<td>Roberts and Loube (case 1)</td>
</tr>
</tbody>
</table>

**Summary**

Forty-three cases of single coronary artery have been reported. This anomaly is thought to be due to absence or misplacement of one coronary artery anlage. In the absence of cardiovascular disease or other anomalies of the heart, single coronary artery is not associated with decreased cardiac function. Autopsy examination of an infant with a single coronary artery usually reveals other anomalies of the heart and great vessels and an atypical distribution of the single vessel. Two additional cases of single coronary artery are reported, one occurring in a white woman 80 years of age.

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