Anomalous Left Coronary Artery Arising from the Pulmonary Artery in an Adult

A Review of the Therapeutic Problem

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ANGIOCARDIOGRAPHY has recently led to the recognition of the anomalous left coronary artery arising from the pulmonary artery in older children and adolescents. Three 16-year old patients are the oldest ones in whom the diagnosis has been made during life.\textsuperscript{1–3} In all of the 14 cases\textsuperscript{4–6} reported from the adult age group (18 years or more) the diagnosis was made at autopsy.

We are reporting here a minimally symptomatic 29-year-old woman with the diagnosis of anomalous left coronary artery established by cineangiocardiography with the electrocardiographic pattern of anterolateral myocardial infarction.

Report of Case

A 29-year-old Caucasian woman was admitted for evaluation of a cardiac murmur and slight left ventricular enlargement. She was known to have had a cardiac murmur since birth. At the age of 3 months, she had prolonged bouts of crying for 1 month, but there was no history of any other distress in infancy. No information was available as to whether or not the crying was aggravated by feeding. During childhood she was slightly more tired on exertion than her peers but could play strenuous games. In school she performed as a drum majorette without difficulty. In her adult life she continued to have slight fatigue on climbing 2 flights of stairs but did not experience any exertional chest pain or other cardiac symptoms.

In June, 1964, she suddenly developed a “fluttering” in her chest while at work as a medical secretary. On climbing 10 stairs, she noticed dyspnea but no chest pain. An electrocardiogram taken at the time showed a supraventricular tachycardia which was resistant to carotid sinus pressure, phenobarbital, or morphine injection. After an hour, gagging was induced, and the arrhythmia reverted to sinus rhythm. She returned to work after a day’s rest and had no further symptoms until December, 1964, when, while she was sitting at her desk, a sharp pain developed at the inferior aspect of her right scapula. It extended to the front of the right side of the chest and into the substernal area. It was made worse by deep breathing but was not oppressive and did not extend to the left arm. It lasted for 4 days and then cleared spontaneously. Because of this episode of chest pain, she was referred for complete evaluation.

On physical examination the heart rate was 80 per minute and regular except for an occasional premature ventricular contraction. Blood pressure was 102/58 mm Hg. The jugular venous pressure was not elevated, and there was no ankle edema. The liver was not palpable. The point of maximal cardiac impulse was located 1 cm to the left of the midclavicular line in the fifth interspace. A minimal left ventricular heave was palpable. Clinically, the second sounds were split in a normal relation to respiration and the pulmonary component was not accentuated. A continuous murmur of grade 2/6 intensity was heard in the second left intercostal space. The systolic component of the murmur became much louder in full expiration.

Serial electrocardiograms over the past 9 years showed left axis deviation, the pattern of old anterolateral myocardial infarction, and peri-infarction block (fig. 1). The cube vectorcardiogram (fig. 2) recorded by Grishman’s method was in accord with these findings. The chest roentgenogram revealed slight enlargement of the left ventricle, normal pulmonary conus, and no increase in the pulmonary vascularity.

Cardiac Catheterization and Cineangiography

Right-heart and left-heart catheterizations were done, and all pressures were normal. Hydrogen
inhalation with a platinum electrode catheter in the pulmonary artery showed a 5.5-second appearance time in contrast to an appearance time in the inferior vena cava of 13 seconds, thus indicating a delayed left-to-right shunt (the usual appearance time in this laboratory for a left-to-right shunt due to septal defect or a patent ductus arteriosus being 1 to 4 seconds). Oximetric determination of oxyhemoglobin revealed an insignificant (0.5 vol/100 ml) rise in the pulmonary artery. A thoracic cineaortogram, performed in the left anterior oblique position, showed a large and tortuous right coronary artery originating from the normal position in the right anterior aortic sinus. There was delayed filling of the left coronary artery through collateral channels from the right coronary artery. The left coronary artery then drained into the pulmonary artery (figs. 3 and 4). The right coronary artery was engaged with the catheter and selective coronary arteriograms were obtained in the right anterior oblique and left anterior oblique projections. The main right coronary artery was 8 mm in diameter, and all of its major branches were dilated and tortuous. Four pathways of collateral filling of the left coronary artery were seen (figs. 3 and 4): (1) A large conus branch of the right coronary artery fed into the anterior descending left coronary artery through Vierusen's ring. (2) Several small vessels over the anterior surface of the right ventricle originating from a marginal branch of the right coronary artery entered the anterior descending left coronary artery midway down its course in the inter-

ventricular sulcus. (3) A network of small vessels coursed through the septum from the greatly enlarged posterior descending right coronary artery to the entire length of the anterior descending left coronary artery. (4) A single small vessel passed from the distal part of the right coronary artery in the left posterior atrioventricular sulcus. A left ventriculogram showed no mitral regurgitation. A pulmonary arteriogram, taken as the patient performed a Valsalva maneuver, showed reflex of contrast material into the ostium of the left coronary artery. Indocyanine-green dye was then injected into the right coronary artery catheter, and samples were drawn from the right atrium, right ventricle, and pulmonary artery. Early appearance in the pulmonary artery confirmed the left-to-right shunt at that level. Normal circulation time was observed following an injection of indicator into the arch of the aorta with the sampling catheter in the pulmonary artery. This demonstrated that the origin of the shunt was in the right coronary artery and not at the site of a ductus arteriosus.

Because the patient had minimal symptoms and was reluctant to undergo any surgical procedures, she was discharged from the hospital to be followed by her physician.

Discussion

Different clinical manifestations have been observed in patients in whom the left coronary artery originates from the pulmonary artery. These differences led Agustsson and
his associates\textsuperscript{7} to classify such cases into two groups. The “adult” type was characterized by intercoronary collateral circulation adequate to permit flow of blood into the left coronary system, and absence of myocardial infarction. The “infantile” type was characterized by paucity of collateral net-

work without demonstrable shunt from the right into the left coronary artery, associated with electrocardiographic pattern of infarction. They suggested that the difference in the anatomy of collaterals was present at birth rather than acquired.

Recent case reports,\textsuperscript{1, 8–10} however, have illustrated that some patients presenting in infancy with signs of severe myocardial insufficiency may show striking clinical improvement and live into childhood. Retrograde thoracic aortography was performed on six of these patients, and all had a shunt from the right into the anomalous left coronary artery. The two children observed by Noren and his associates\textsuperscript{10} exhibited marked regression of the electrocardiographic pattern of anterolateral myocardial infarction which had been present in infancy. Our patient shows that adult life with minimal symptoms may be achieved even in the presence of the pattern of transmural anterolateral myocardial infarction. Well-developed intercoronary collateral vessels have been demonstrated in infants\textsuperscript{11, 12} who had clinical findings typical of the infantile type.

These cases are not consistent with the concept of distinct functional types among patients with anomalous origin of the left coronary artery and lend support to the recently advanced theory of changing phases\textsuperscript{13} in a given patient. According to this concept, a transition exists for a period after birth between the phase of forward flow from the pulmonary artery into the anomalous left coronary artery and the phase of well-developed intercoronary anastomosis permitting retrograde flow in pulmonary artery. During the transitional phase, perfusion of the portion of the myocardium supplied by the anomalous artery is almost certainly inadequate and severe ischemia or death may result.

The tachycardia which brought our patient to the attention of a physician is of interest because arrhythmias may be a cause of the sudden death common in the adult group.\textsuperscript{4}
Retrograde cineaortogram, left anterior oblique view, demonstrating retrograde flow through the anomalous left coronary artery. A, left. Diastole, 1.27 seconds after injection in the aortic root. A dilated and tortuous coronary artery arises from the right aortic sinus. There is no filling of the left coronary artery at this time, B, right. Diastole, 3.15 seconds after injection. The right coronary is enlarged throughout its course. A heavy "blush" is seen in the region of the ventricular septum, representing small collateral vessels coursing from the posterior descending to the anterior descending artery through the interventricular septum. The left coronary artery (arrow) is now seen, and the pulmonary artery has opacified. Motion analysis of this sequence and the selective coronary arteriograms in the left and right oblique projections permitted construction of a composite diagram (fig. 4).

Surgical Treatment

In a small postmortem series of older patients (age 16 to 60 years) with anomalous left coronary origin, the mean age of death was 35 years. The majority died suddenly, having been asymptomatic and not known to have heart disease. These statistics render the current patient's benign clinical course less reassuring and prompted a review of the current status of surgery in this condition.

Ligation of the anomalous left coronary artery at the origin has been the most successful and widely used procedure, although experience has been confined largely to infants. Of 27 patients who underwent ligation, 16 survived (table 1), and 13 improved either by reduction in heart size, or by improvement in the electrocardiogram. The nonsurvivors were all infants; of the eight on whom clinical information was available, seven had definitive evidence of myocardial infarction (table 1). The oldest patient operated on, aged 17 years, developed ischemic electrocardiographic changes after the operation.

The preoperative demonstration of retrograde flow through the anomalous left coronary artery into the pulmonary artery assures...
Table 1

Consequences of Ligation of Anomalous Left Coronary Artery in Twenty-Seven Patients

<table>
<thead>
<tr>
<th>Number and age</th>
<th>16 Survivors</th>
<th>11 Dead*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>9 Infants (1-22 mo)</td>
<td>6 Children (2-12 yr)</td>
</tr>
<tr>
<td>ECG</td>
<td>M1—9</td>
<td>LVH—5</td>
</tr>
<tr>
<td>No clinical data available</td>
<td></td>
<td></td>
</tr>
<tr>
<td>L→R shunt present</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>No preoperative determination</td>
<td></td>
<td></td>
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<tr>
<td>Follow-up</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean time</td>
<td>16.7 mo</td>
<td>10.4 mo</td>
</tr>
<tr>
<td>Clinical improvement</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>No change</td>
<td>2</td>
<td>ECG after ligation</td>
</tr>
</tbody>
</table>

MI, myocardial infarction; LVH, left ventricular hypertrophy.

*Personal communication from R. D. Bowe.
†Personal communication from C. S. Weldon.
‡Two deaths were due largely to technical difficulties at operation.

the best chance of survival following ligation. Fifteen of the 16 survivors had demonstrable left-to-right shunts, while only four of the nonsurvivors had documented retrograde flow (table 1). Associated mitral regurgitation, when severe, may portend a poor operative prognosis, since the valvular dysfunction probably is evidence of severe myocardial damage and papillary muscle dysfunction.

Corrective surgery, with implantation of the anomalous left coronary artery into the aorta or anastomosis with a systemic vessel is theoretically a more desirable procedure because the aim is to establish a more normal two-coronary pattern. The limited experience with this operation, however, did not justify its recommendation to the asymptomatic patient whose case is presented herein. The uniqueness of the current case and the adverse electrocardiographic change following ligation in the reported 17-year-old patient weighed heavily in the decision to postpone surgery for the present.

Summary

The 29-year-old woman with anomalous left coronary artery arising from the pulmonary trunk whose case is presented is at the time of writing, the oldest patient on record in whom this condition has been diagnosed during life. The diagnosis was made by aortic cineangiography and confirmed by selective injection of contrast material into the right coronary artery. The collateral pathways between the right and the anomalous left coronary artery were thus defined. Vectorcardiographic and electrocardiographic patterns of anterolateral myocardial infarction with perinfarction block have not been observed in any of the previously reported adults with this type of rare congenital anomaly.

The 27 cases of ligation of the aberrant left coronary artery arising from the pulmonary artery reported in the literature have been reviewed and the results of this operation discussed.

Diagnosis of the anomalous left coronary artery in adult life is possible if it is considered in patients presenting with a continuous murmur at the base and electrocardiographic evidence of myocardial infarction.

Acknowledgments

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