Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

Surgical Treatment by Ostial Occlusion Through Pulmonary Arteriotomy

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SUMMARY
Two cases of anomalous origin of the left coronary artery from the pulmonary artery are reported in asymptomatic patients 15 and 27 years of age, bringing to 30 the total number of cases of this anomaly reported in the postinfantile age group. ECG exercise tests demonstrated marked ischemic changes in both patients, but angina or abnormal shortness of breath did not develop in either patient during or after exercise. The anomalous vessel was interrupted by closure of its ostium through a pulmonary arteriotomy during cardiopulmonary bypass, a procedure felt to offer a distinct technical advantage in the surgical treatment of this condition. Following ligation of the anomalous vessel, ECG exercise tests no longer showed definitive evidence of myocardial ischemia. This is interpreted as evidence that ligation should reduce the incidence of exercise-induced sudden deaths in this anomaly.

Additional Indexing Words:
Congenital heart disease Angina Ligation of anomalous coronary artery
ECG exercise test Myocardial ischemia Left-to-right shunt
Sudden death

Anomalous origin of the left coronary artery from the pulmonary artery in patients beyond infancy has received increased attention since Lampe and Verheugt and Agustsson and associates showed that this abnormality could be demonstrated during life by angiography. Patients beyond 2 years of age with this anomaly are usually asymptomatic, but the high reported incidence of sudden death has prompted consideration of several techniques for surgical treatment. The demonstrated retrograde blood flow in the anomalous vessel has suggested that ligation at its origin from the pulmonary artery would be beneficial and this has been the procedure used in most cases in which surgery has been performed. Anastomosis of the anomalous vessel to the aorta has been successfully accomplished in two cases reported by Cooley and co-workers. Although ligation is logical therapy, follow-up has been too brief to assess its effect on the incidence of sudden death. If symptoms were present, they usually resolved postoperatively, a striking example of this being reported by Baue and associates. The electrocardiogram
has usually been unchanged after operation, but two reports\textsuperscript{11, 12} have described further abnormalities after surgery.

Two cases of this anomaly in young adults have been diagnosed at this institution, and the abnormal vessel has been interrupted by use of a new surgical approach. The exercise electrocardiogram before and after surgery was used to demonstrate the beneficial effect of ligation on myocardial ischemia.

**Report of Cases**

**Case 1**

C. S., a 15-year-old white male, was referred in November 1966, because of a heart murmur discovered on routine examination. He was completely asymptomatic and participated in vigorous physical exercise without difficulty. His infancy was normal, and there was no family history of heart disease. He presented as a healthy, well-developed 15-year-old male. The blood pressure was 120/50 mm Hg and the pulse was 80 per minute. The lungs were clear. The jugular venous pulse was normal and the carotid pulse was quick rising and of normal amplitude. A mild left ventricular heave was present in the fifth intercostal space at the midclavicular line. The first and second heart sounds were normal. A grade III/VI, harsh systolic ejection murmur was heard from the aortic area to the apex, and a grade II/VI, diastolic blowing murmur, along the right and left sternal borders. The murmurs were continuous in the pulmonic area. On the remainder of the examination findings were normal. Blood and urine were normal on routine examinations, as were chest roentgenograms. The ECG showed Q waves in leads I and aVL and voltage criteria for left ventricular hypertrophy (fig. 1). An ECG exercise test showed marked ischemic S-T depression (fig. 2), although the patient had no symptoms during the test. Cardiac catheterization demonstrated a small increase in oxygen saturation of the pulmonary arterial blood but otherwise disclosed normal conditions (table 1). Supravalvular aortic angiography showed a large right coronary artery filling sequentially multiple

![Figure 1](http://circ.ahajournals.org/)

*Figure 1*

Patient C.S., resting electrocardiograms. The postoperative records show a rightward and anterior shift of the T-wave axis, and decrease in QRS voltage.
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Figure 2

Patient C.S., ECG exercise tests. Marked ischemic S-T depression occurs immediately after exercise before, and 1 month after, surgery. Four months after surgery only J point depression is present.

collateral vessels, the left coronary artery, and finally the pulmonary artery (fig. 3).

On July 7, 1967, surgery was performed through a median sternotomy, using standard cardiopulmonary bypass techniques. The right coronary artery was huge and tortuous and communicated via multiple large collateral vessels with the left coronary system. The left coronary artery arose from the posterior sinus of the pulmonary artery and bifurcated approximately 1.5 cm from its origin into left anterior descending and circumflex branches. It was extremely thin-walled and appreciably softer than the right coronary. Systolic and diastolic thrills were palpable at its origin. No areas of gross myocardial scarring were visible.

The orifice of the anomalous left coronary artery was approached through a short pulmonary arteriotomy. During systole, arterialized blood spurted from the left coronary artery into the pulmonary artery, necessitating aortic cross-clamping. Local myocardial cooling with iced saline and elective ventricular fibrillation were employed. The anomalous orifice, measuring approximately 0.5 by 0.5 cm, was then easily closed in two layers, and security of the closure was tested by release of the aortic clamp and defibrillation. The aorta was cross-clamped for a total of 10 minutes. Following bypass, the left coronary artery became much more distended and tense; there were no residual thrills. No significant arrhythmias occurred.

The postoperative course was uneventful, without significant enzyme changes, and the patient was discharged on the seventh postoperative day.

Postoperatively the heart sounds were normal and no significant murmurs were noted. The patient remained asymptomatic with excellent exercise tolerance as before surgery. ECG exercise tests were performed 1 and 4 months postoperatively at a slightly higher exercise level than before surgery (fig. 2).

Case 2

S. S., a 27-year-old white woman, was referred for evaluation of a heart murmur in February
Patient C.S. Cineangiogram at 48 frames per second following supravalvular aortic injection. (A) Frame 15. A large right coronary artery is seen arising from the aorta normally. The left coronary artery is not visualized. (B) Frame 52. The right coronary artery is still visible (black arrow, left) and in addition the left coronary (black arrow, right) and pulmonary (white arrow) arteries are opacified.

Table 1

<table>
<thead>
<tr>
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<th>C.S. (case 1)</th>
<th>S.S. (case 2)</th>
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Abbreviations: RA = right atrium; RV = right ventricle; PA = pulmonary artery; PAW = pulmonary artery wedge; BA = brachial artery; m = mean; s/ed = systolic/end-diastolic; s/d = systolic/diastolic.

1967. At age 7, she was found to have an enlarged heart. When she was 11 years old, heart murmurs were first heard, and rheumatic heart disease with mitral insufficiency, patent ductus arteriosus, and ventricular septal defect were suspected. At 23, pregnancy and full-term delivery were uncomplicated. The patient’s development had been unremarkable, and she had been asymptomatic except for mild fatigability throughout her life.
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Figure 4

Patient S.S., resting electrocardiograms. The postoperative changes are similar to those of patient C.S.

Physical examination showed a well-developed, healthy-appearing young woman with blood pressure of 110/60 mm Hg and pulse rate of 76 per minute. The lungs were clear. The jugular venous pulse showed a prominent a wave with normal pressure, and the carotid pulse was quick rising. A mild left ventricular heave was felt 2 cm lateral to the midclavicular line in the fourth intercostal space. A systolic thrill was present from the lower left sternal border to the apex. The first heart sound was slightly increased at the apex. The second heart sound was slightly increased along the left sternal border and was normally split during inspiration. A grade III/VI continuous murmur was best heard at the apex, the systolic component extending well to the left sternal border and the diastolic component well laterally to the midaxillary line. On the remainder of the examination results were normal. Blood and urine were normal on routine examinations. Chest roentgenograms showed a slightly increased heart size with prominent pulmonary vasculature and aortic knob. A resting ECG (fig. 4) showed left ventricular hypertrophy and prominent Q waves in leads I and aV L. ECG exercise test showed marked and prolonged ischemic S-T depression (fig. 5), although the patient was asymptomatic throughout exercise. Cardiac catheterization demonstrated a left-to-right shunt at the level of the pulmonary artery, but other findings were
normal (table 1). A supravalvular aortic angiogram showed a somewhat enlarged ascending aorta and a huge tortuous right coronary artery with multiple large collateral vessels supplying the left coronary artery which emptied into the pulmonary artery (fig. 6).

On November 3, 1967, surgery was performed, as in case 1, through a median sternotomy and use of cardiopulmonary bypass. Again the right coronary artery was large and tortuous and communicated through multiple channels with the left coronary artery which arose from the posterior pulmonary sinus. The left coronary artery branched immediately beyond its origin. The anomalous orifice measured approximately 1.5 by 1.0 cm and was closed without difficulty through an anterior pulmonary arteriotomy (fig. 7). The aorta was cross-clamped for 12 minutes during this portion of the procedure to control retrograde flow through the anomalous vessel.

Again, following closure, pressure within the left coronary artery increased remarkably and there were no residual thrills.

The postoperative course was uncomplicated, and the patient was discharged on the ninth postoperative day.

One month after operation, the patient was asymptomatic. Findings on examination were normal, except for a grade II/VI, scratchy, probably extra-cardiac systolic murmur localized to the base. A repeat ECG exercise test at an identical exercise level showed no definite evidence for ischemia (fig. 4).

Discussion

Gouley and later Agustsson and associates emphasized two different modes of presentation of anomalous origin of the left coronary artery from the pulmonary artery which

Figure 5

Patient S.S., ECG exercise tests. Before surgery, marked ischemic S-T depression is present, and it persisted for 15 minutes after exercise was stopped. After surgery, definite ischemic changes do not occur.
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Figure 6

Patient S.S. Lateral view angiogram with 6 exposures per second. (Left) Exposure 3. A hugely dilated and tortuous right coronary artery arises normally from the aorta with massive collaterals at the apex. The left coronary artery is not visualized. (Right) Exposure 7. There is continued visualization of the right coronary artery (curved arrow) with filling of the left coronary artery (arrow, mid-right) and pulmonary artery (straight arrows, upper left).

they designated as “infantile” and “adult” types. “Postinfantile” is a more appropriate designation than “adult,” as nearly all patients over 2 years of age have the clinical presentation described for the “adult” type. The infantile condition is characterized by episodes of crying and pallor, gross cardiomegaly, congestive heart failure, and myocardial infarction, with death before age 1 year. Mitral insufficiency is a common associated lesion, presumably due to papillary muscle infarction or dysfunction. In marked contrast, the postinfantile patient with this anomaly usually has no symptoms during infancy and remains asymptomatic in later years or has only mild fatigability or dyspnea with vigorous exertion. Anginal pain is infrequent. An apical heave with systolic and often diastolic heart murmurs is present and is suggestive of aortic insufficiency, patent ductus arteriosus, or ventricular septal defect. The ECG usually shows a leftward axis, prominent Q waves in leads I and aV L, abnormal T waves, and voltage criteria for left ventricular hypertrophy which may be most prominent in the limb leads. Cardiac catheterization has shown no abnormality or a small increase in oxygen saturation in the pulmonary artery. The diagnosis is made by angiography. Although a compatible murmur is present in half the cases, significant mitral insufficiency is uncommon. It has, however, been reported as the dominant clinical feature in three patients. With antemortem diagnosis being possible, a third category of patients has become apparent; that is, an infant with congestive heart failure survives and improves, and by the age of 3 or 4 years is

Circulation, Volume XXXVIII, July 1968
asymptomatic or nearly so.\textsuperscript{4, 6, 15, 16, 18} This group has led Edwards\textsuperscript{19} to emphasize that the condition is a continuum from inadequate to adequate intercoronary collaterals, and to favor abandonment of the concept of types. We feel that there is indeed a continuum, but that the concept of infantile and postinfantile types is useful both prognostically and therapeutically. To the present time, including our cases, 30 cases of the postinfantile type of this anomaly have been reported in the literature.\textsuperscript{1–3, 5, 10–13, 17, 20–24} Sixteen of these were diagnosed at postmortem examination.\textsuperscript{3, 5, 17} Sudden death had occurred in 12 of the 16; in eight it had occurred without prior symptoms. Death was frequently related to vigorous exercise and occurred most often in the third or fourth decade of life. The oldest reported patient was a woman who died an accidental death at age 60.

Brooks\textsuperscript{25} reported the first cases of this anomaly in 1886. From the autopsy finding of a veinlike left coronary artery and his knowledge of the relative pressures in the aorta and pulmonary artery, he concluded that the left coronary flow was retrograde. This opinion was overlooked, and in subsequent reports the myocardial abnormalities were ascribed to perfusion of the left coronary
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system with venous blood. Case and associates\(^8\) and Edwards,\(^9\) in 1958, renewed the concept of fully oxygenated blood flowing from the normally placed right coronary through collaterals into the left coronary system and then into the pulmonary artery. This was proven at surgery,\(^{26}\) then by angiography,\(^2\) and has since been demonstrated repeatedly by angiography and at cardiac catheterization, where an increase in blood oxygen saturation may be found in the pulmonary artery. Perfusion of the myocardium with desaturated blood cannot therefore be invoked as the cause of the observed abnormalities. The left-to-right shunt is usually small, in contrast to our case 2, and cannot be considered large enough to increase left ventricular work significantly. It is believed that the low pressure in the left coronary artery, which is close to that in the pulmonary artery, results in low perfusion pressure at the capillary level and inadequate tissue oxygenation, particularly during the stress of exercise.

The surgical treatment of anomalous left coronary artery has recently been reviewed by Cooley and associates.\(^6\) In the postinfantile type, the only acceptable alternatives are ligation of the anomalous vessel at its origin, transposition to the aorta by a graft, or primary anastomosis. Each of these approaches involves a tedious and occasionally dangerous dissection of the left main coronary artery which is usually extremely thin-walled and easily torn. Dissection may be especially difficult if the coronary artery lies partially within the myocardium or if the bifurcation occurs close to the origin. The creation of a “two-coronary” system by anastomosis of the anomalous vessel to the aorta is esthetically pleasing, but carries no certain physiological benefit. As noted by Smith,\(^{27}\) the anomaly of single coronary artery is compatible with a normal life span. It is reasonable to assume that the single coronary system resulting from interruption at its origin of an anomalous left coronary artery, subserved by abundant large collaterals, is capable of supplying full physiological needs. Some assurance of this may be taken from the operative finding of increased perfusion pressure in the left coronary system, equal to that in the right, following closure of the fistulous tract.

The operative approach used in our cases, that is, closure of the anomalous coronary artery from within the pulmonary artery under cardiopulmonary bypass, provides full control and obviates the need for dissection of the delicate anomalous vessel, and is felt to offer a distinct technical advantage in the surgical approach to this condition.

Ligation has been performed and reported in 14 patients over 2 years of age with this anomaly without mortality.\(^2, 4, 10–12, 15, 21, 22, 24\) Those patients who had symptoms were significantly improved. The effect on the life expectancy cannot be assessed at this time but will require long-term follow-up. Some authors have reported reversion of the postligation resting ECG toward normal.\(^2, 15, 24\) but most have reported no change. Others have reported development of further abnormalities\(^{11, 12}\) and have questioned the efficacy and safety of the procedure.\(^{16, 20}\)

ECG exercise tests have not been described in previous reports. Both of our patients performed tests before and after surgery (figs. 2 and 5). Preoperatively both patients showed marked exercise-induced ischemic S-T depressions. Abnormalities lasted 15 minutes after cessation of exercise in case 2. Neither patient had any symptoms other than the normal amount of tachypnea during or after exercise. Resting T-wave abnormalities similar to those reported by Massih and co-workers\(^{11}\) developed in both patients following surgery, and could be due to myocardial damage or injury. However, these changes may as well be due to postoperative epicardial changes associated with pericardiomy, or to changes in repolarization caused by an increase in the pressure in the massive intercoronary artery collateral system and the capillaries of the left ventricle. The ECG exercise test 1

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month postoperatively in case 1 showed persistence of an ischemic response to exercise, but 4 months after surgery only J point depression was seen and definite ischemic changes were absent. The 1-month postoperative exercise test in case 2 showed no definite evidence of ischemia, in marked contrast to the profound and prolonged ischemia of the preoperative record.

We feel that the ECG exercise response gives substantial evidence that ligation of the anomalous artery at its origin in the post-infantile patient is beneficial and can be expected to reduce the incidence of exercise-induced sudden death. The explanation for the delayed reversal of exercise ischemia in case 1 is not clear, but it is possible that the capillary bed of the left coronary artery is underdeveloped because of the low pressure in that system, and that its expansion may require a period of weeks or months after the pressure is increased. The absence of angina in these patients, in the presence of clear-cut electrocardiographic evidence of myocardial ischemia, is a curious and unexplained feature. Theories on the mechanism of anginal pain production should undertake to explain this.

References


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Circulation. 1968;38:113-123
doi: 10.1161/01.CIR.38.1.113
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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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:
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