Single Left Coronary Artery with Fistula to Right Ventricle

Reconstruction of Two-Coronary System with Dacron Graft

By Grady L. Hallman, M.D., Denton A. Cooley, M.D., Dan G. McNamara, M.D., and Joseph R. Latson, M.D.

Anomalies of origin and distribution of coronary arteries are unusual malformations; they may occur in multiple forms and may produce striking physical findings and sometimes serious cardiac disability. In one anomaly the entire coronary arterial blood supply is provided by a single vessel rather than by the customary two. Coronary artery-right ventricular fistula is another malformation that is well recognized and frequently produces findings similar to arteriovenous fistula. Recently we treated a patient who had a single left coronary artery combined with coronary artery-right ventricular fistula. The purpose of this report is to describe the diagnostic procedures employed and to demonstrate a definitive technic for surgical treatment, which corrected both aspects of the anomaly.

Case Report

The patient was an 8-year-old girl admitted on December 17, 1963, with a history of fatigue on effort beginning in infancy. Three weeks before admission exertional fatigue and weakness had become more severe and she had noted epigastric pain while running.

Physical examination revealed a small girl underweight for her age. No venous distention or arterial pulsation was noted in the neck. The apex beat was in the sixth intercostal space in the midclavicular line and there were a forceful precordial thrust and a systolic thrill at the left sternal border. Long, high-pitched systolic and diastolic murmurs were heard along the left sternal border. There was such a brief pause between the two that the murmurs at first appeared to be continuous. Unlike the typical murmur of patent ductus arteriosus, however, the diastolic component was high-pitched rather than low-pitched and was not decrescendo. The volume of the peripheral pulses was slightly increased and the blood pressure was 100/50. There were no additional abnormalities on physical examination.

Roentgenogram of the chest revealed no significant increase in the transverse diameter of the heart (fig. 1). The right atrial shadow was prominent and the left cardiac border was blunt, suggesting left ventricular hypertrophy. The pulmonary vascular markings were slightly increased.

The electrocardiogram was interpreted as indicating left ventricular hypertrophy because of flattening of the ST segment in precordial leads

Figure 1
Anteroposterior roentgenogram of chest prior to operation showing cardiac size at upper limits of normal. Pulmonary vascular markings are increased.

From the Cora and Webb Mading Department of Surgery and the Department of Pediatrics, Baylor University College of Medicine and the Junior League Clinic of the Texas Children’s Hospital, Houston, Texas.

Supported in part by the U. S. Public Health Service (HE-05387) and (HE-03137), the Houston Heart Association, and the Institute for Aid and Rehabilitation, Houston, Texas.

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V₆ and V₇. The QRS-T angle in the frontal plane was + 120 degrees.

The diagnoses considered included patent ductus arteriosus, coronary artery-right ventricular fistula, ventricular septal defect with aortic or pulmonic regurgitation, and aortic septal defect.

Cardiac catheterization under local analgesia revealed an increase in oxygen saturation in the right ventricular outflow tract. The pulmonary-to-systemic flow ratio was 1.54:1. Pressures in the right heart were normal. A retrograde arterial catheter was placed in the ascending aorta and radiopaque dye was injected (figs. 2 and 3). A dilated left coronary artery was seen. The circumflex branch of this vessel passed to the right and anteriorly, ultimately following the course normally occupied by the right coronary artery. This anomalous vessel terminated in the outflow tract of the right ventricle. No right coronary artery was visualized.

Operation was performed with the patient in the supine position. A median sternotomy incision was made, and the pericardium was incised vertically. A single large left coronary artery arose in the usual position from the aorta (fig. 4). At the left border of the pulmonary artery it divided into anterior descending and circumflex branches. The circumflex branch was tortuous and dilated, 6 to 8 mm. in diameter, and passed around the

**Figure 2**
Left. Representative frame of angiocardiogram with catheter in ascending aorta (anteroposterior view). Contrast material outlines a single large left coronary artery whose circumflex branch continues in the distribution of a right coronary artery and ends in the outflow tract of the right ventricle. Right. Composite drawing of angiocardiographic findings (anteroposterior view).

**Figure 3**
Left. Representative frame of angiocardiogram with catheter in ascending aorta (lateral view). Right. Composite drawing of angiocardiographic findings (lateral view).
posterior surface of the heart in the atroventricular groove, where it gave off a posterior descending branch. It then continued onto the anterior surface of the heart between the right atrium and ventricle in the groove normally occupied by the right coronary artery. This vessel terminated in the outflow tract of the right ventricle, where a continuous thrill could be felt. Dissection was begun at this terminal portion of the coronary artery, and its point of entrance into the right ventricle was ligated with 2-0 silk. A vascular clamp was placed 1 cm. proximal to this point, and the artery was transected near the fistula. A partial occluding vascular clamp was placed tangentially to the anterior surface of the aorta 1 cm. above the annulus, excluding a portion of aortic wall. An incision was made in the excluded aorta, and a 6-mm. knitted Dacron tube graft with its proximal end beveled at 45 degrees was anastomosed end-to-side to the aorta with 5-0 polyethylene suture. The graft was then cut at a length of 3 cm. and its distal end was anastomosed end-to-end to the anomalous coronary artery with the same suture material. Vascular clamps were removed and pulsatile flow was established into the coronary artery from the aorta. The thoracic incision was closed in layers with retrosternal tube drainage. The postoperative course was uneventful, and the continuous murmur was no longer audible. The patient was discharged on the seventh postoperative day. She was re-admitted 6 weeks later by which time she had returned to full activity and was asymptomatic. Follow-up cardiac catheterization and angiography revealed that the left-to-right shunt had been eliminated and that the aortocoronary graft was patent (fig. 5).

**Discussion**

Murray\(^1\) reported two patients with a single coronary artery and fistulous communication with the right ventricle. Recovery followed ligation of the fistula. Reviewing the literature (1963) he found 72 cases of single coronary artery and 105 with coronary artery fistula. There had been only two cases reported previously with both lesions.\(^2,3\) In the same year Michaud et al.\(^4\) described three patients with coronary artery-right ventricular fistulas, one of whom had a single left coronary artery. All three patients survived ligation of the fistula.

One of the earliest and most comprehensive reviews of the subject of single coronary artery was published in 1950 by Smith,\(^5\) who collected 43 cases from the literature and added two of his own. None had fistulas. He classified patients with this anomaly into three categories depending on the distribution of the single coronary artery: (1) those whose single vessel followed the course of only one coronary artery, (11 cases); (2) those in whom the single artery was present in the distribution of both coronary arteries (18 cases); and (3) those in whom the distribution of the vessel was atypical (15 cases). Infants usually fell in category 3 and had additional anomalies of the heart and great vessels. In the 45 cases review by Smith\(^5\)

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**Figure 4**

*Drawings of findings at operation and repair utilized. (Upper left). Midline sternotomy incision. Single large left coronary artery gives off anterior descending branch and continues as dilated tortuous circumflex artery. Circumflex artery travels in the atroventricular groove and continues onto anterior surface of heart in position normally occupied by right coronary artery, ending in fistula to outflow tract of right ventricle. (Upper right). Right ventricular portion of anomalous coronary artery dissected out, distal end ligated, and vascular clamp applied. (Lower left). Coronary artery divided. Incision made in anterior surface of aorta within confines of partial occluding vascular clamp. (Lower center). Six-millimeter Dacron tube graft anastomosed to aorta and to coronary artery. (Lower right). Clamp removed and flow established from aorta into anomalous coronary artery via graft.*

*Circulation, Volume XXXII, August 1965*
single coronary artery was associated with symptoms only if additional cardiovascular disease or other anomalies of the heart were present.

The principal hazard of a single coronary artery is emphasized by Roberts and Loube in a report on nine patients with this anomaly, three of whom died from myocardial infarction. They found that patients with only one coronary artery may remain asymptomatic during early life, but later are especially vulnerable to the effects of arteriosclerotic occlusive lesions, since they have only one source of arterial inflow into their coronary systems. An effort was made to protect the patient in the present report from this hazard by establishing a dual coronary supply. This was accomplished by anastomosing the right ventricular portion of the coronary artery to the aorta with use of a Dacron tube graft. Furthermore, demonstration that such a procedure is feasible may recommend similar technics for the correction of anomalous origin of the coronary artery from the pulmonary artery. Current practice in the treatment of such defects calls for ligation of the anomalous vessel, a procedure that leaves the patient with a single source of arterial inflow into the coronary arterial system and is fraught with danger in infants with poorly developed collateral circulation.

The surgical treatment of coronary arterial fistulas is now well established. Since the first successful operation detailed by Biork and Crafoord in 1947, many reports and reviews have been published on the diagnosis and treatment of this anomaly. There is general agreement that closure of the fistula is desirable even in patients without severe symptoms. Maintenance of continuity of the involved vessel after closure of the fistula is considered an important principle of surgical technic. Simple ligation of the communication in the patient in this report would have sufficed to interrupt the left-to-right shunt from the aorta to the right ventricle, but would have left only one source of inflow into the coronary arterial system, namely, the ostium of the left coronary artery. Anastomosis of the right ventricular portion of the coronary artery to the aorta via a Dacron graft restored a dual coronary supply and should protect against the hazards of arteriosclerotic occlusive lesions in later life.

**Summary**

The case of an 8-year-old girl with a single
coronary artery is presented. This vessel arose as a left coronary artery and followed the distribution of one coronary artery. The long circumflex branch traveled in the atrioventricular groove and continued as a vessel occupying the position usually held by the right coronary artery. It ended as a fistula to the outflow tract of the right ventricle. A continuous murmur was present clinically. Cardiac catheterization and selective angiography demonstrated the anatomy of the lesion and the presence of a left-to-right shunt. Operation consisted of ligation of the fistula and anastomosis of the right ventricular portion of the coronary artery to the aorta with a Dacron tube graft. This latter maneuver established a dual inflow into the coronary arterial system and should overcome the disadvantage of a single coronary artery in the event that arteriosclerotic occlusive lesions develop in later life. Furthermore, the procedure demonstrated the feasibility of grafting technics that may find application in the treatment of anomalous origin of a coronary artery from the pulmonary artery.

References
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_Circulation_. 1965;32:293-297
doi: 10.1161/01.CIR.32.2.293

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1965 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/32/2/293

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