Atresia of the Left Coronary Artery Ostium

Repair by Saphenous Vein Graft

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SUMMARY
Symptoms and electrocardiographic changes of acute myocardial ischemia in a 10-year-old boy resulted from congenital atresia of the ostium of the left coronary artery. At 14 years of age persistence of symptoms and a positive exercise test prompted surgical treatment by aortocoronary artery saphenous vein bypass graft. Two months postoperatively, the symptoms and the abnormal findings on exercise test had subsided and forward flow through the graft to the left coronary artery was demonstrated by contrast angiography.

Additional Indexing Words:
Two coronary artery systems Single coronary artery Treadmill exercise

CONGENITAL malformations of the coronary arteries, though uncommon, deserve special emphasis because of the potential benefit by surgical treatment. In this center, aortocoronary artery bypass using a Dacron tube was used first by Dr. Grady Hallman on December 23, 1963,1 to establish two coronary systems in a patient who had single left coronary artery with fistulous connection to the right ventricle. The success of this procedure prompted the use of saphenous vein grafts for repair of anomalous origin of the left coronary artery from the pulmonary artery.2-4 This report presents a child who suffered from myocardial ischemia due to atresia of the left coronary artery ostium. The distal left coronary artery and its branches were normal in size and distribution, though filling occurred only by collaterals from the right coronary artery system. The patient was successfully treated by an aorto-coronary artery-saphenous vein bypass in February, 1972, in Texas Children’s Hospital.

Case Report
A male patient, 10 years of age, had been healthy except for the incidental finding of a mitral systolic murmur at 5 years of age. A few months before referral to this center, he developed a fainting episode while sitting as a spectator at a school athletic event. He was referred to this center because while playing basketball he again experienced syncope for a few seconds followed by severe retrosternal pressure-like pain radiating to both arms, shortness of breath, and diaphoresis. Family history revealed no other incidence of known congenital heart disease or early coronary artery disease. On admission to Texas Children’s Hospital, the heart rate was 90 beats/ min with occasional irregularity due to premature ventricular contractions; blood pressure was 110/80, and there were no manifestations of peripheral vascular disease. The heart sounds were of normal intensity. There was a Grade II/VI late systolic murmur and a third heart sound at the apex. The serum cholesterol, total lipids, triglycerides, lipoprotein pattern,
serum enzymes (SGOT, CPK, and LD), and erythrocyte sedimentation rate were normal. Electrocardiograms showed anteroseptal injury and ischemia which cleared in a few days. The heart size and contour were normal as evidenced by X-rays. With a clinical impression of hypertrophic obstructive cardiomyopathy, the patient was catheterized. The left coronary artery filled through collaterals from the right coronary artery and the proximal segment of the main left coronary artery was not visualized by serial film angiography with only aortic root injection. No selective coronary cineangiograms were done during this investigation. Left ventricular angiocardiography demonstrated mild mitral insufficiency. At that time, the impression was obstructive disease of the main left coronary artery of unknown etiology and pathologic anatomy.

Between 11 and 14 years of age, the patient was complaining of palpitations, due to the premature ventricular beats, and shortness of breath on moderate exertion. Treatment included propranolol and isosorbide dinitrate with reduction of the premature ventricular contractions.

In January, 1972, at 14 years of age, his

**Figure 1**

The electrocardiogram which was recorded immediately postexercise shows 3-mm straight S-T segment depression in V5 and V6.

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condition was reevaluated. The symptomatology and physical findings were the same. The previously mentioned laboratory investigations were repeated and remained normal. The resting electrocardiogram was normal but the patient developed 3-mm straight S-T segment depression with increase of the heart rate from 75 to 125 beats/min after 7 min of exercise (3 min at 1.7 mph and 4 min at 3 mph at 10% elevation) on a treadmill (fig. 1). Selective coronary (fig. 2) and aortic root cineangiography using the femoral approach demonstrated filling of the left coronary system from the right coronary artery through collaterals. The proximal main left coronary artery segment was never well visualized and it was impossible to selectively enter the left coronary artery with the catheter. Apart from the proximal segment, the retrogradely filled left coronary system appeared to be normal. Narrowing was seen in the proximal right coronary artery and was thought to be due to spasm, although a proximal disease similar to the one involving the left coronary artery was considered. No filling of the pulmonary artery was seen either after the aortic root or the selective right coronary artery injections. This study demonstrated that the patient had complete occlusion of the proximal left coronary artery.

On February 10, 1972, the patient underwent operation. There was no anomalous origin of left coronary artery. Direct observation through aortotomy revealed that the proximal right coronary artery was normal. A blind dimple was seen at the site of the left coronary ostium. A biopsy was taken from the aortic wall above the left coronary ostium and was found to be histologically normal. The wall of the opened left anterior descending coronary artery was normal. An aorto-left anterior descending coronary artery-saphenous vein bypass graft was inserted.

Postoperatively, the patient had an uneventful recovery. The shortness of breath and the arrhythmia disappeared, but the Grade II/VI late systolic apical murmur was still heard. Treadmill exercise testing and coronary cineangiography by the Sones technic were repeated 2 months postoperatively. The patient was able to run 10 min (3 min at 1.7 mph, 2 min at 3 mph, 2 min at

Figure 2

Two preoperative views of the right coronary artery (right anterior oblique and left anterior oblique) and a composite drawing of angiocardiographic findings (anteroposterior view) showing retrograde filling of the normal left coronary artery branches from collaterals, a persistent narrowing of the proximal right coronary artery, and a large right coronary artery.
4 mph, and 3 min at 5 mph all at 10% elevation) with increase of the heart rate from 72 to 185 beats/min without any ischemic changes. The coronary cineangiograms showed that the left coronary artery filled only from the patent saphenous graft (fig. 3). No narrowing in the proximal right coronary artery and no filling of the left coronary system from the right coronary artery were seen.

**Discussion**

The anatomy of the described anomaly, the absence of associated cardiac defects, and the relatively old age differentiates the condition in our patient from single coronary artery malformations\(^6,7\) and from the several varieties of coronary occlusive disease in infancy.\(^8-12\) Pre- and postoperative angiocardio graphic studies (figs. 2, 3), as well as surgical exploration, showed that the described patient has two coronary arterial systems. The preoperative retrograde filling of the left coronary system through collaterals from the right coronary artery differentiates our patient from the expected antegrade filling of single coronary artery branches. The collateral vessels are

**Figure 3**

Postoperative right coronary (posteroanterior view), bypass injections (right anterior oblique view), and a composite drawing of angiocardio graphic findings (anteroposterior view) showing that the normally distributed left coronary artery fills only from the patent graft, that there is no spasm in the proximal right coronary artery, and that the size of the right coronary artery has decreased.
similar to those described in acquired left coronary artery obstruction\textsuperscript{18} and anomalous origin of left coronary artery from the pulmonary artery.\textsuperscript{14} Ogden\textsuperscript{15} used the term "absence of a proximal coronary artery associated with a single ostium in the other aortic sinus"—a suitable description for the anomaly reported here—but Ogden’s diagram and description illustrates a single coronary artery.

Chest pain, syncope, shortness of breath, arrhythmias, resting, and postexercise ECG changes indicated that our patient had myocardial ischemia. This was confirmed by the disappearance postoperatively of symptoms and by the objective increase in his exercise tolerance. The myocardial ischemia, despite abundant collateral circulation and normal-size left coronary artery, indicates that collaterals alone were inadequate for perfusion. The lesion, in fact, may provide us with a naturally occurring model to assess what might be expected for the patient with anomalous origin of the left coronary artery from the pulmonary artery who is treated by surgical ligation of the left coronary artery as it leaves the pulmonary artery.

The anatomic appearance of the lesion by direct vision at operation plus the normal histology of the aortic wall biopsy suggests that the abnormality is a developmental defect. According to Grant,\textsuperscript{16} the first embryonic indication of coronary arteries appear as thickenings of the aortic endothelium just before division of the truncus communis into aorta and pulmonary trunk. The arterial rudiments are at first solid columns of cells which later acquire a lumen and grow outward into the superficial portion of the myocardium. Assuming that the lesion reported is a developmental defect, the mechanism may be failure of canalization of the proximal segment of the left coronary anlage. Hudson\textsuperscript{17} reported a similar anomaly in an infant who had died at 8 months of age in which the left coronary artery ended in an area of fibrosis 5 mm from its origin with a very small distal left coronary artery, and felt that the left coronary anlage failed to connect with its artery.

This report emphasizes that myocardial ischemic symptoms at a young age may be due to congenital coronary artery anomaly and may benefit from coronary cineangiography and saphenous vein bypass operation.

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References


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