Isotopic Findings in Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery: Report of an Adult Case

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Anomalous origin of the left coronary artery from the main pulmonary trunk results in myocardial ischemia or infarction, and may be a cause of death in the first months of life. Some patients, however, develop satisfactory coronary collateral circulation and remain asymptomatic into adulthood. In these patients, myocardial perfusion and left ventricular function are not well understood. We report the case of a 17-yr-old female patient, suffering from anomalous origin of the left coronary artery from the main pulmonary trunk, who underwent reimplantation of the left coronary artery to the aorta. The preoperative permanent ^{201}Tl defect of the left antero-lateral ventricular wall and the abnormal regional wall motion induced by stress exercise testing were fully reversed after the operation.

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Anomalous origin of the left coronary artery from the main pulmonary trunk is the most frequent of the congenital anomalies of the coronary arteries. During fetal life, pressure regimens and oxygen saturation of the aorta and of the pulmonary artery are not much different from one another so that no significant ischemia results from the perfusion of left ventricular myocardium by blood originating from the pulmonary circulation. After birth, the drop of oxygen saturation and the pressure regimen in the pulmonary circulation results in sudden ischemia, which may lead to the development of myocardial infarction and subsequent left ventricular function deterioration. In some patients, however, a collateral circulation develops, preventing severe myocardial ischemia and serious myocardial damage. In these rare cases, survival until adulthood is possible.

We report one such case where isotopic investigations of myocardial perfusion and left ventricular performance played an important role in the decision-making process.

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CASE HISTORY

A 17-yr-old female patient without history of chest pain during exertion was referred to our institution after a cardiac murmur was detected. Her weight and height were normal and she was asymptomatic. A continuous murmur could be heard on the 3rd left interspace. No other abnormality was apparent. Cardiothoracic ratio was 0.55. The pattern of electrocardiogram indicated an anteroseptal infarction. Two-dimensional echocardiography was normal, but the left main coronary stem could not be seen arising from the aorta; Doppler was not available at this time.

Exercise stress testing conducted on a bicycle ergometer demonstrated inadequate blood pressure evolution during exercise with systolic blood pressure tending to remain at its baseline levels. No significant ST changes appeared despite the fact that heart rate was increased to 90% of maximal age-predicted heart rate. Except for some degree of dizziness at peak exercise no symptoms occurred.

During selective coronary arteriography, the left coronary artery could not be detected arising from the aorta. A massive collateral network, however, was seen arising from the right coronary artery which filled the left coronary system. The main pulmonary trunk was injected through the left main stem arising from it. Catheterization of right and left heart chambers showed a normal pressure and a minor left-to-right shunt at the main pulmonary trunk level. Left ventriculography showed a normal contraction pattern. Ejection fraction was 0.57.

The patient was referred for surgery, which consisted of direct reimplantation of the left main stem back to the aorta. The electrocardiogram did not change during the immediate postoperative course, which was uneventful. Four and a half years later, the patient is alive and asymptomatic. R-waves obtained at rest were nearly normal in leads V1 to V3. A two-dimensional echocardiogram shows enlarged left and right coronary arteries normally arising from the aorta.

Radionuclide Study

Thallium-201 SPECT. Three microcuries of ^{201}Tl-chloride were injected in an antecubital vein at peak exercise. Data acquisition began 5 min after exercise. Four hours later a redistribution study was recorded.

Radionuclide Angiography. Multigated equilibrium blood-pool imaging was performed using ^{99}Tc-labeled red cells. Data were obtained at rest and at peak exercise with the gamma camera detector in a 35° left anterior oblique position.

Preoperative ^{201}Tl SPECT demonstrated an antero-lateral defect at peak exercise. During the redistribution study, the antero-

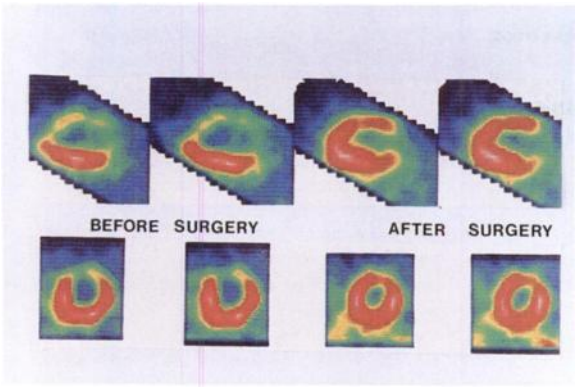

FIGURE 1. Pre- and postoperative ^{201}Tl SPECT. Sagittal slices are shown on the upper row and coronal slices on the lower. Reduction uptake is seen in the septum and anterior wall before surgery. Postoperative exercise scintigram shows persistent high activity in the same area.

lateral defect persisted, suggesting permanent scarring (Fig. 1). On radionuclide ventriculography, left ventricular ejection fraction (LVEF) was normal at rest (0.59) but failed to increase during exercise because of a significant decrease of regional LVEF in the septal and apical area contrasting with a normal increase of regional LVEF on the lateral wall (Table 1).

Postoperative rest-SPECT study showed marked improvement over the preoperative one. One year later, repeated SPECT showed normal perfusion of the left myocardium at rest as well as at peak exercise (Fig. 1). LVEF was normal at rest and rose normally at peak exercise without significant anomaly in regional wall motion 2 yr after surgery (Table 1).

DISCUSSION

The Bland-Garland-White syndrome is a rare congenital anomaly observed in 1 out of 400 patients suffering from congenital heart disease (*2,4,5*).

The diagnostic utility of ^{201}Tl SPECT has been previously reported by several authors (*2–4*). Anterior wall thallium defects are usually observed in this disease. Moddie, who reported three cases of Bland-Garland-White syndrome in adults (*5*), showed that anterior wall thallium defects were observed preoperatively in 3 patients; postoperatively, thallium scintigraphy returned to normal at rest as well as at peak exercise in only one patient who had no history of myocardial infarction; conversely, in the two remaining patients, the thallium defect did not disappear after surgery. In our patient, a large thallium defect of the antero-lateral wall of the left ventricle was observed preoperatively at rest as well as at peak exercise. This aspect of irreversible thallium defect is usually related to the presence of scar tissue due to myocardial infarction. It should be noted that the distribution study was performed 4 hr after exercise. If some late redistribution occurred, it might have been visualized on a third acquisition 24 hr later.

Global and regional wall motion was studied by using bidimensional echocardiography in infants suffering from severe left ventricular impairment during the first months of life, but not in adults. In our opinion, the assessment of left ventricular function may be of interest in predicting the result of surgical repair. In our patient, no anomaly in the regional wall motion was apparent on resting radionuclide angiography which became abnormal only during exercise. The lack of increase in global LVEF at peak exercise was due to a depression of the regional ejection fraction in the septal and antero-apical area. This fact is usually interpreted as the sign of ischemic myocardium during exercise rather than as scar tissue from myocardial infarction. The normalization of myocardial perfusion and of regional and global LVEF during exercise postoperatively confirmed this notion. In our patient, we may suppose that the ischemic myocardium was prevented

TABLE 1
Regional Wall Motion on ^{99m}Tc Ventriculography

Regional EF	Rest		Exercise		Variation	
	*	+	*	+	*	+
Septum	0.63	0.53	0.51	0.59	−0.12	0.06
	0.64	0.52	0.44	0.62	−0.20	0.10
	0.60	0.57	0.46	0.66	−0.14	0.09
	0.58	0.65	0.56	0.70	−0.02	0.05
	0.55	0.65	0.54	0.73	−0.01	0.08
Apex	0.55	0.63	0.55	0.80	0	0.17
	0.60	0.69	0.55	0.84	0.01	0.15
	0.62	0.68	0.65	0.82	0.03	0.14
Lateral	0.62	0.70	0.75	0.77	0.13	0.07
wall	0.57	0.67	0.65	0.74	0.12	0.07
	0.57	0.62	0.68	0.70	0.11	0.08
	0.58	0.53	0.69	0.57	0.11	0.04
LVEF	0.58	0.60	0.56	0.66	−0.02	0.06

* Preoperative regional ejection fraction.

† Postoperative regional ejection fraction.

Preoperatively, LV contraction appears normal at rest, but exercise induces marked hypokinesis in the septum and apical area. Two years after reimplantation of the left coronary artery, maximal exercise test does not induce any wall motion abnormality.

from definitive myocardial necrosis due to an abundant collateral circulation provided by the right coronary system.

The practical concept that emerges is that in the Bland-Garland-White syndrome the observation of a thallium defect at rest and at peak exercise does not necessarily imply that the defect area consists of scar tissue nor does it necessarily mean that irreversible lesions have occurred in the area of the abnormally originating artery. The study of global and regional wall motion may be useful for predicting the result of surgical repair of the defect (i.e., reimplantation of the left coronary artery back to the aorta).

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