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Anomalous left main coronary artery arising from the pulmonary artery in an adult: Treatment by internal mammary artery grafting

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Left main coronary artery arising from the pulmonary artery is reported to occur once in every 300,000 live births and accounts for 0.25% to 0.5% of congenital heart disease cases. Without treatment, most infants die during the first year of life. The small number who survive infancy presumably live until adulthood because of good collateral blood flow. Reports of surgical correction in adults are scarce. We present a case of this anomaly in an adult whose myocardial ischemia and left ventricular dilatation were completely reversed by use of the left internal mammary artery as a conduit for revascularization of the left coronary artery system after ligation of the origin of the anomalous left main artery from the pulmonary artery.

A 42-year-old woman sought treatment for fatigue and exertional dyspnea. Examination revealed precordial systolic and diastolic murmurs. Electrocardiography showed left ventricular hypertrophy. Transesophageal echocardiography showed a dilated left ventricle (diastolic dimension 6.2 cm) and aneurysmal dilatation of the coronary arteries. The left main coronary artery orifice could not be

identified in the left coronary sinus of Valsalva. Cardiac catheterization revealed a right-to-left shunt of 1.5 to 1. Angiography showed a markedly dilated right coronary artery with delayed collateral filling of the left coronary artery system, which drained retrogradely into the pulmonary artery via an anomalous left main coronary artery (Fig. 1). Exercise testing showed poor exercise tolerance, hypotension, and widespread ischemic changes. Thallium 201 perfusion scintigraphy demonstrated a large reversible anteroapical defect (Fig. 2).

In view of the clinical findings, surgical correction was recommended. During cardiopulmonary bypass, it was impossible to arrest the heart because of brisk oxygenated blood flow into the left coronary system from noncoronary collaterals. The left main coronary artery orifice in the pulmonary artery was identified posterolaterally on the left side and was oversewn from within and ligated on the outside of the pulmonary artery. The left internal mammary artery was anastomosed to the left anterior descending artery as a pedicle graft.

Three months after the operation, she was free of symptoms and had excellent exercise tolerance (9 minutes on the Bruce treadmill protocol compared with 4 minutes before the operation). Stress perfusion scintigraphy revealed uniform myocardial perfusion (Fig. 2). Echocardiography showed normal left ventricular size (diastolic dimension 5.5 cm). Doppler echocardiography demonstrated brisk antegrade diastolic flow down the left internal mammary artery.

An anomalous left main coronary artery arising from the pulmonary artery is a rare finding in adults. The demonstrated presence of coronary and noncoronary collateral blood flow in this case presumably contributed to the woman's survival beyond childhood. Because of the association with sudden death and myocardial ischemia, surgical correction is usually recommended once the diagnosis is established. However, the optimal surgical technique for adults with this coronary anomaly remains unclear. Possible surgical techniques include the following: (1) ligation of the left main artery with revascularization of the left coronary system using saphenous vein aorta-coronary bypass grafting¹ or internal mammary artery grafting, which has been reported in only one case²; (2) reimplantation of the left main artery to the aorta directly or indirectly through a pulmonary artery tunnel³ or via an internal iliac artery graft.⁴ Saphenous vein grafts are well recognized to have poor long-term patency rates. Direct reimplantation of the left main artery to the aorta is often not technically possible because of unfavorable anatomy. Intrapulmonary tunneling has been associated with pulmonary valvular dysfunction.

The case presented here demonstrates that brisk antegrade blood flow and an excellent functional outcome can be obtained with the left internal mammary artery used as the conduit for revascularization of the left coronary system after its separation from the pulmonary artery. This is contrary to a previous suggestion that the difference in diameter between the mammary artery and the dilated left coronary artery system may cause competitive blood flow resulting in graft occlusion.⁴ Because of the likelihood of long-term patency, the left internal mam-

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Fig. 1. Selective contrast angiography in the right anterior oblique view showing the markedly dilated and tortuous right coronary artery (RCA) and filling of the left coronary arteries via collateral vessels.

Fig. 2. Thallium 201 perfusion scintigraphy in the vertical long-axis view at stress (A) and after redistribution (B), demonstrating a reversible anteroapical defect. After the operation, stress (C) and redistribution (D) images showed uniform perfusion.

mary artery should be the conduit of choice in adult patients with this coronary anomaly undergoing corrective operations.

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