

sis. It is doubtful that inadequate alpha-receptor blockade was a significant factor, since the blood pressure was well controlled with phenoxybenzamine. Although T wave changes may occur in patients with mitral valve prolapse⁹ (a condition suggested by this patient's initial physical examination), it seems unlikely that this would explain the close association between ECG changes and specific interventions (i.e., drug therapy, tumor excision). Failure of the ECG to make a complete return to normal 4 months postoperatively suggests that minor irreversible subendocardial necrosis may have occurred, but these ECG changes may also be related to mitral valve prolapse.⁹

In conclusion, this case demonstrates that diverse ECG abnormalities of rhythm, conduction, and repolarization may occur in patients with pheochromocytoma. The paroxysmal nature and different degree of catecholamine secretion from this tumor probably accounts for the variability of these changes. Importantly, the ECG abnormalities may persist or worsen following adequate alpha-adrenergic blockade. This may occur in the absence of obstructive coronary artery disease or other clinically detectable cardiac pathology. Although the mechanism for these changes is unclear, unopposed beta-adrenergic receptor stimulation may be a major factor. The nonspecificity and acute nature of the ECG changes associated with pheochromocytoma will often warrant coronary angiography prior to surgery. Although alpha-adrenergic blockade and blood pressure control are essential before any invasive procedure, complete alpha- and beta-receptor blockade may be important for further myocardial protection, especially when ECG abnormalities are present initially.

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Successful treatment of angina pectoris with liver transplantation and bilateral internal mammary bypass graft surgery in familial hypercholesterolemia

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Familial hypercholesterolemia (FH) is a disease characterized by elevated plasma cholesterol levels, tendon xanthomas, xanthelasmas, and widespread atherosclerosis resulting in premature acute myocardial infarction and sudden cardiac death.¹ The severity of atherosclerosis is directly related to the plasma low-density lipoprotein (LDL) cholesterol level. Previous attempts to treat coronary artery disease in homozygous patients have been unrewarding. With a persistent elevation in plasma cholesterol concentration, the atherosclerotic process continues unabated. Surgical management of angina in these patients has been similarly unrewarding; saphenous vein bypass grafting carries the risk of dislodging and embolizing atherosclerotic plaques from the ascending aorta at the time of operation, since the atherosclerosis is particularly severe in the ascending aorta at the site of graft anastomosis.² Moreover, a persistent elevation in plasma cholesterol is associated with a high rate of saphenous vein graft occlusion. Percutaneous transluminal coronary angioplasty has also been disappointing in patients with homozygous FH due to a high frequency of early restenosis (unpublished observations). We report a patient with severe FH who has had successful alleviation of angina pectoris and who has maintained graft patency 1 year following liver transplantation and bilateral internal mammary artery (IMA) bypass grafting. This is the first report of combined liver transplantation and IMA implantation as a radical treatment approach for this form of advanced coronary artery disease.

VP is a 13-year-old young man who had xanthomas detected at age 18 months that led to the diagnosis of homozygous FH. He was treated with a low-fat, low-cholesterol diet until age 5, when cholestyramine was started; at age 9, nicotinic acid was added. When the patient was referred to the National Institutes of Health (NIH) at age 11, he complained of exertional angina. He had widespread yellow xanthomas, a 3/6 systolic ejection-type murmur at the upper sternal border, and bruits over both carotid arteries and both femoral arteries. He was

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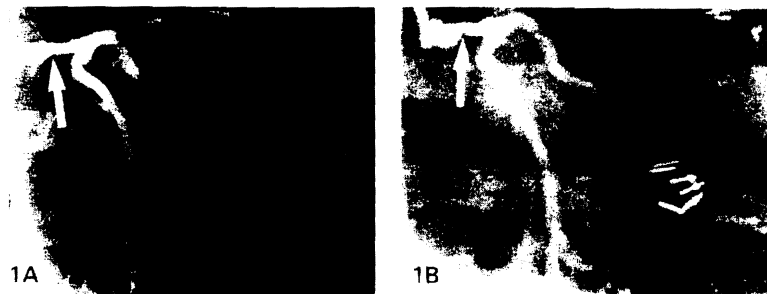


Fig. 1. Angiogram of the left coronary artery in the right anterior oblique projection before (A) and after (B) IMA implantation, showing no progression of atherosclerosis in the left main coronary artery (arrow). B shows the circumflex and obtuse marginal arteries with the left anterior descending artery filling with nonopacified blood via the IMA graft.

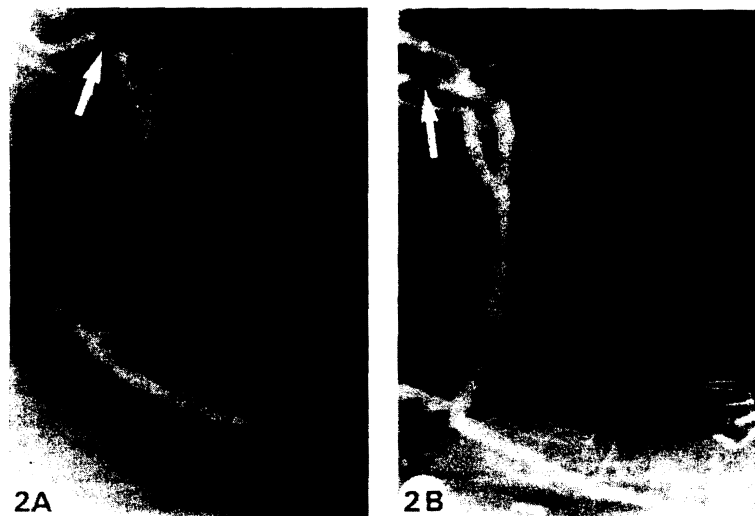


Fig. 2. Angiogram of the right coronary artery in the right anterior oblique projection before (A) and after (B) IMA implantation, showing no progression of atherosclerosis at the ostium (arrow) of the right coronary artery.

treated with combinations of cholestyramine, nicotinic acid, and lovastatin, but he had continued angina and sustained a right parietal cerebrovascular accident at age 11. Cardiac catheterization showed severe stenosis of the ostia of both the left main and right coronary arteries (Figs. 1, A and 2, A). There was an atherosclerotic narrowing of the ascending aorta at the sinotubular level but no pressure gradient across this narrowing. Radionuclide angiography showed a resting ejection fraction of 64% at rest, which fell to 46% during upright bicycle exercise, indicating significant inducible myocardial ischemia.

He was started on biweekly plasma exchanges and underwent liver transplantation at Children's Hospital of Pittsburgh on November 6, 1985. Postoperatively, he sustained a non-Q wave myocardial infarction complicated by transient pulmonary edema. Because of continued angina, he underwent bypass grafting at the Cleve-

land Clinic on January 27, 1986. The operation consisted of a left IMA anastomosis to the left anterior descending artery and a right IMA anastomosis to the right coronary artery. His postoperative course was uneventful.

Following liver transplantation, total cholesterol decreased from 1170 to 254 mg/dl, LDL cholesterol decreased from 1062 to 152 mg/dl, and high-density lipoprotein (HDL) cholesterol increased from 22 to 43 mg/dl.³ With these changes, there has been a marked reduction in the xanthomas. Following coronary bypass grafting, there has also been a marked resolution of the patient's angina. He exercised for 20 minutes with the NIH combined protocol (stage 8 = 3.1 mph, 16% grade) without ischemic ECG changes, and radionuclide angiography showed an ejection fraction of 56% at rest, which increased normally to 64% during upright bicycle exercise. Thirteen months following bypass graft surgery, he underwent repeat cardiac catheterization. Both internal

mammary grafts were widely patent. Angiography of the native coronary artery showed no progression of disease (Figs. 1, B and 2, B).

Homozygous FH is a rare disorder usually resulting in death in the second or third decade of life due to advanced proximal coronary disease. The extraordinary interventions in this patient have produced gratifying short-term results and provide important insights that may be applicable to patients with coronary artery disease and less severe forms of hypercholesterolemia.

It appears that correcting the underlying metabolic abnormality through liver transplantation has diminished the manifestations of severe hypercholesterolemia. Since transplanted hepatic tissue is rich in LDL receptors,^{3,4} this procedure produced a dramatic fall in plasma cholesterol, (76% in this patient), which resulted in a marked reduction of xanthomas and halted the progression of coronary atherosclerosis.

In addition, this patient illustrates the utility of IMA bypass grafting.⁵ After 1 year follow-up, this patient has no apparent narrowing of the IMA grafts and is free of angina. Compared to other patients with homozygous FH who have undergone saphenous vein bypass graft surgery, this patient appears to have a good chance of long-term amelioration of angina and survival.

Finally, a significant reduction in LDL cholesterol levels may lead to improved patency of grafts after surgery and may halt the progression of atherosclerosis in patients with less striking hypercholesterolemia. Blankenhorn et al.⁶ demonstrated that reducing LDL cholesterol levels with combined bile acid sequestrant-niacin treatment resulted in significantly less progression of coronary artery disease. Therefore the findings in the present case may have broader implications. In summary, we report the first example of successful combined liver transplantation and IMA bypass grafting as a treatment of angina pectoris in a patient with homozygous FH.

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Giant right atrial thrombus causing right ventricular inflow and outflow obstruction

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Recent reports¹⁻⁶ have indicated that two-dimensional echocardiography (2DE) is the investigation of choice for detecting right-sided intracavitary thrombi. We report a patient in whom 2DE and angiography demonstrated an extremely large, right atrial (RA) mass that proved on operation to be a pedunculated, organized thrombus.

A 25-year-old woman presented with a 1-month history of palpitation, dizziness, and a syncopal attack. Her past history was unremarkable. On physical examination, she was in no acute distress. Her blood pressure was 110/70 mm Hg, with a pulsus paradox of 15 mm Hg, her pulse was 120 beats/min and regular, and the respiratory rate was 16/min. Her neck veins were fully distended. Cardiac examination was remarkable for a left parasternal heave, a grade II/VI holosystolic murmur, a short early diastolic murmur varying in intensity with postural changes, an atrial gallop, and to and fro scratchy sounds, all heard along the left parasternal border. The liver was pulsatile 4 cm below the right costal margin but there was no peripheral edema. Homans' sign was negative and there was no calf tenderness. Routine laboratory tests, including arterial blood gases, were unremarkable except for low voltage and right axis deviation on electrocardiography. 2DE demonstrated a large, tongue-shaped, noncalcified RA mass, which although fixed to the RA septum, protruded through the tricuspid valve and remained in the RA and right ventricular cavities throughout the cardiac cycle (Fig. 1). The left ventricle was pushed by a markedly dilated right ventricle. The inferior vena cava appeared free of abnormal echoes. Cardiac catheterization was performed with great caution. Mean RA pressure was 19 mm Hg, while right ventricle and pulmonary artery pressures were normal. Right heart cineangiography confirmed the 2DE findings and showed the obstructing mass lodged in the right ventricular outflow tract (Fig. 2). The main pulmonary artery and the proximal portions of its subdivisions were normal. Lung scan was negative for pulmonary embolization (PE). The diagnosis of RA myx-

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