Total arterial revascularization in a child with familial homozygous hypercholesterolemia

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Premature coronary artery occlusive disease in familial homozygous hypercholesterolemia might necessitate coronary bypass surgery in children and young adults. Arterial grafts have been shown to be superior to venous grafts in children, with longer patency rates and a better growth potential.^{1,2} Young adults with familial hypercholesterolemia also fare better with internal thoracic grafts, although the benefit of multiple arterial grafts is still unproved in this setting.^{3,4} Surgical angioplasty of the main coronary arteries is another alternative to preserve the precious arterial conduits for future reoperations.⁵ Saphenous venous grafts, although inferior, have also been found to have some growth potential and long-term patency, even up to 22 years.^{6,7}

The subject of this report is a 12-year-old child with familial homozygous hypercholesterolemia and severe obstructive coronary artery disease who underwent a total arterial revascularization. To the best of our knowledge, this is the first report of total arterial revascularization at this age.

Clinical Summary

A 12-year-old boy (weight, 22 kg) presented to our cardiac clinic with a history of exertional angina of more than 2 years' duration. His anginal symptoms had shown recent worsening with frequent rest pains, and at present, he was unable to walk even 100 m without becoming symptomatic. He had a strong family history of hypercholesterolemia, premature coronary artery disease, and death but had no other classic major risk factors. He had multiple tendinous xanthomas over his fingers, elbow and knee joints, and Achilles tendon. He was being treated with statins, but the lipid profile was grossly deranged, with a total cholesterol level of 670 mg/dL, low-density lipoprotein (LDL) cholesterol level of 600 mg/dL, high-density lipoprotein cholesterol level of 39 mg/dL, and triglyceride level of 136 mg/dL. Electrocardiography showed reversible ST-T depression in lateral leads. Echocardiographic Doppler scanning revealed mild aortic regurgitation and trivial mitral regurgitation. Coronary angiography showed severe left main and proximal right coronary artery disease (Figure 1). He was treated with intravenous nitroglycerin, β -blockers, angiotensin-converting enzyme inhibitors, and aspirin, along with lipid-lowering drugs.

He was taken up for myocardial revascularization. The bilateral internal thoracic artery (ITA; pedicle graft) and the left radial artery (free graft) were harvested. The left ITA/right ITA measured 1 mm, and the radial artery measured 1.5 mm. During cardiopulmonary bypass and cardioplegic arrest, the left anterior descending artery and the right coronary artery were revascularized with the left and right ITA, respectively, and the obtuse marginal artery was revascularized with the radial artery. The child became asymptomatic after coronary artery bypass grafting (CABG), and a cineangiogram on the 12th postoperative day showed patent arterial grafts with no anastomotic problems (Figure 2).

The child was started on bezafibrate and cholestyramine (INN: colestyramine) in addition to atorvastatin. Plasmapheresis and LDL apheresis could not be carried out because of the

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Received for publication January 28, 2004; revisions received March 11, 2004; accepted for publication March 15, 2004

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J Thorac Cardiovasc Surg 2005;129:932-4
0022-5223/\$30.00

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doi:10.1016/j.jtcvs.2004.03.054

Figure 1. Aortic root injection in the left anterior oblique view showing a tight left main coronary artery (LMCA) lesion and a proximal right coronary artery (RCA) lesion.

high recurrent cost of these procedures, which the family could not afford. Similarly, partial liver transplantation was not considered because of financial reasons and also because this facility is as yet not fully developed in our country. Lipid control has remained suboptimal, with LDL cholesterol still greater than 300 mg%. The child has been free of angina over the past 1 year.

Discussion

Familial hypercholesterolemia is an autosomal dominant disorder in which mutation in the gene encoding the LDL receptor causes a high plasma level of cholesterol, with accelerated atherosclerosis and development of multiple tendon xanthomas. Although the common age of presentation is late in the second and third decade, earlier presentation with severe obstructive coronary artery disease is well documented.⁸ The youngest child reported to have undergone CABG for familial hypercholesterolemia is a 7-year-old in whom the left ITA measured only 1 mm, and venous grafts were made to the left anterior descending artery and the right coronary artery.⁸ The benefits of arterial grafts in older patients with familial hypercholesterolemia have been documented in several studies in terms of actuarial survival and freedom from symptoms.^{3,4} Aggressive lipid lowering with LDL apheresis and plasmapheresis is extremely important to prevent recurrence, and new lesions occur if total cholesterol remains greater than 220 mg%.^{9,10}

The use of arterial conduits for coronary problems in infants and children is now well accepted, with excellent long-term patency and growth in children with Kawasaki disease, postarterial switch, and other congenital coronary lesions.^{1,2} Differences in adaptation to growth of children have been shown between arterial and venous grafts, with thoracic artery growing in proportion to somatic growth, whereas saphenous vein grafts tend to course in a more linear way, with no increase in length or diameter.¹¹ However, isolated reports have highlighted excellent growth and patency of venous grafts up to 22 years after bypass surgery.^{6,7} Surgical coronary angioplasty of the left main artery has also been shown to be effective in the midterm and is useful in avoiding venous and arterial conduits and preserving these for later reoperations.⁵

Figure 2. A, Postoperative angiogram showing a patent left internal thoracic artery (LIMA) to left anterior descending artery (LAD) anastomosis. B, Postoperative angiogram showing a patent right internal thoracic artery (RIMA) to right coronary artery (RCA) anastomosis. C, Postoperative angiogram showing a patent left radial artery to obtuse marginal artery anastomosis.

In our patient, a 12-year-old boy with left main disease and disease of the proximal right coronary artery, we revascularized

the vessels with total arterial grafts, despite the technical difficulties of harvesting and anastomosing the conduits. Whether multiple arterial grafts in this setting provide a better long-term outcome is not known from the available limited literature.³ The gastroepiploic artery has been used as the additional arterial conduit in patients with Kawasaki disease, but we are not aware of the use of the radial artery in children.¹ We preferred the radial artery because of its adequate size and length and also because of its being an arterial conduit. We chose to do a total arterial revascularization, expecting better long-term patency, especially when aggressive lipid lowering was not possible, and our patient can hardly afford an early redo operation that would be more likely with venous conduits. The patency of autologous saphenous vein in children is not satisfactory. The patency of the vein graft is $65.4\% \pm 7.9\%$ 84 months after the operation in those of more than 8 years of age but only $27.7\% \pm 8.1\%$ ($P < .01$) in those younger than 7 years of age.¹² Furthermore, surgical expertise in performing CABG in adults and neonatal arterial switch allowed the concerned surgeon (AB) to be confident of a good anastomotic result.

In conclusion, we describe total arterial revascularization in a young boy with familial hypercholesterolemia. Whether such a strategy in the absence of measures like LDL apheresis and plasmapheresis can be better than venous grafts is not known. In developing countries, in which financial difficulties prevent routine use of such expensive treatment regimens, this could be an alternative that allows better long-term outcome.

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