Open Journal of Cardiovascular Surgery

CASE REPORT

Triple Vessel Coronary Artery Bypass Grafting in a 14-year-old Child with Familial Hypercholesterolemia-A Rare Case Report

S Borkar Shirish[1], S Ganesh Kamath[1], Sagar C.V. Sunil[2], Bedjirgi Chidanand[3] and Kashyap Nitin[3]

[1]Consultant Cardiovascular and Thoracic Surgery. [2]Consultant Cardiac Anesthesia. [3]Senior Registrar Cardiovascular and Thoracic Surgery, Department of Cardiovascular Surgery, Kasturba Medical College, Manipal University.
Email: drshirishborkar@hotmail.com

Abstract: Familial hypercholesterolemia is a genetic disorder caused by a mutation in the low density lipoprotein (LDL) receptor gene. The homozygous type of the disease is rare and causes tendon xanthomas and coronary artery disease during the early years of life. Premature coronary artery occlusive disease in familial homozygous hypercholesterolemia might necessitate coronary bypass surgery in children and young adults We present the case of a 14-year-old boy with familial hypercholesterolemia and coronary artery disease. He underwent triple-vessel coronary artery bypass grafting with bilateral pedicled internal mammary artery and saphenous vein grafting without adverse events. Pediatric patients with familial hypercholesterolemia may present with premature coronary atherosclerosis requiring coronary artery bypass grafting. In situ internal mammary artery grafts should be the graft of choice. To the best of our knowledge, he is one of the youngest such patients reported in the English-language literature who underwent coronary artery bypass surgery.

Keywords: famiilial hypercholesterolemia, triple vessels disease, CABG

Open Journal of Cardiovascular Surgery 2009:2 11–13

This article is available from http://www.la-press.com.

Case Report

A 14-year-old boy (weight, 34 kg) presented to our cardiac clinic with a history of exertional angina of more than 3 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. Patient was diagnosed as type III familial dysbetalipoproteinemia with defect in synthesis in Apo-lipoprotienE_2 after genetic workup. He had arcus juvenilis and multiple tendinous xanthomas over his fingers, elbow, buttocks. He was being treated with statins, as the lipid profile was grossly deranged, with a total cholesterol level of 457 mg/dL, low-density lipoprotein (LDL) cholesterol level of 411 mg/dL, high-density lipoprotein cholesterol level of 29 mg/dL, and triglyceride level of 81 mg/dL before starting statins treatment. Duplex of bilateral carotid and vertebral arteries showed 65% diffuse circumferential wall thickening of the left common carotid artery with hypoplastic right vertebral artery. Electrocardiography showed reversible ST-T depression in lateral leads. Coronary angiography showed severe stenosis of distal left main (90%), mid LAD (80%), proximal right coronary artery disease (ostial 90%) and OM_1 (osteoproximal 90%). He was treated with sorbitrate, β-blockers and aspirin, along with lipid-lowering drugs. He was taken up for myocardial revascularization. The bilateral internal thoracic artery (ITA; pedicle graft) and the saphenous vein graft were harvested. The left ITA/right ITA measured 1 mm, and the saphenous vein measured 1.5 mm. Size of LAD, RCA and OM_1 were 1.75, 2.0, and 0.5 mm respectively. 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 saphenous vein graft. The child became asymptomatic after coronary artery bypass grafting (CABG). The child was started on Ezetimebe in addition to atorvastatin. Plasmapheresis and LDL apheresis could not be carried out because of the high recurrent cost of these procedures, which the family could not afford Lipid control has remained suboptimal, with LDL cholesterol still greater than 300 mg %.

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.[1] 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.[1] 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.[2,3] 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%.[4,5]

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.[6,7] 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.[8] However, isolated reports have highlighted excellent growth and patency of venous grafts up to 22 years after bypass surgery.[9,10] 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.[11]

In our patient, a 14-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.[2] 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.[6] We preferred the saphenous vein because of its adequate size and length. We chose to do triple vessel coronary revascularization, expecting better long-term patency, especially when aggressive lipid lowering was not possible, and our patient can hardly afford an early redo operation. The patency of autologous saphenous vein in children is satisfactory. The patency of the vein graft is 65.4% ± 7.9% 84 months after the operation in those of more than 8 years of age but only 27.7% ± 8.1% ($P < 0.01$) in those younger than 7 years of age.[12] In conclusion, we describe triple vessel coronary 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 any other option 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.

Disclosures

This manuscript has been read and approved by all authors. This paper is unique and is not under consideration by any other publication and has not been published elsewhere.

References

1. Ersoy U, Guvener M. Coronary revascularization in seven-year-old boy with homozygous familial hypercholesterolaemia. *Acta Paediatr*. 2000;89:1501–2.
2. Kawasuji M, Sakakibara N, Fujii S, Yasuda T, Watanabe Y. Coronary artery bypass surgery with arterial grafts in familial hypercholesterolemia. *J Thorac Cardiovasc Surg*. 2000;119:1008–13.
3. Takahashi T, Nakano S, Shimazaki Y, Kaneko M, Hirata N, Nakamura T, et al. Long-term appraisal of coronary bypass operations in familial hypercholesterolemia. *Ann Thorac Surg*. 1993;56:499–505.
4. Fukuzawa S, Ozawa S, Inagaki M, Morooka S, Inoue T. Secondary prevention with lipid lowering therapy in familial hypercholesterolemiaa correlation between new evolution of stenotic lesion and achieved cholesterol levels after revascularization procedures. *Intern Med*. 1999;38:330–5.
5. Richter WO, Donner MG, Hofling B, Schwandt P. Long-term effect of low-density lipoprotein apheresis on plasma lipoproteins and coronary heart disease in native vessels and coronary bypass in severe heterozygous familial hypercholesterolemia. *Metabolism*. 1998;47:863–8.
6. Yoshikawa Y, Yagihara T, Kameda Y, Taniguchi S, Tsuda E, Kawahira Y, et al. Result of surgical treatments in patients with coronary-arterial obstructive disease after Kawasaki disease. *Eur J Cardiothorac Surg*. 2000;17:515–9.
7. Mavroudis C, Backer CL, Ouffy CE, Pahl E, Wax OF. Pediatric coronary artery bypass for Kawasaki congenital, post arterial switch, and iatrogenic lesions. *Ann Thorac Surg*. 1999;68:506–12.
8. Kameda Y, Kitamura S, Taniguchi S, Kawata T, Mizuguchi K, Nishioka H, et al. Differences in adaptation to growth of children between internal thoracic artery and saphenous vein coronary bypass grafts. *J Cardiovasc Surg (Torino)*. 2001;42:9–16.
9. El-Khouri HM, Oanilowicz OA, Slovis AJ, Colvin SB, Artman M. Saphenous vein graft growth 13 years after coronary bypass in a child with Kawasaki disease. *Ann Thorac Surg*. 1998;65:1127–30.
10. Suda Y, Takeuchi Y, Ban T, Ichikawa S, Higashita R. Twenty-two-year follow-up of saphenous vein grafts in pediatric Kawasaki disease. *Ann Thorac Surg*. 2000;70:1706–8.
11. Bonnet O, Bonhoeffer P, Sidi O, Kachaner J, Acar P, Villain E, et al. Surgical angioplasty of the main coronary arteries in children. *J Thorac Cardiovasc Surg*. 1999;117:352–7.
12. Kitamura S, Kameda Y, Seki T, Kawachi K, Endo M, Takeuchi Y, et al. Long term outcome of myocardial revascularization in patients with Kawasaki coronary artery disease. *J Thorac Cardiovasc Surg*. 1994;107:663–74.