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CASE REPORT

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Coronary artery bypass grafting in a 12-year-old girl with familial hypercholesterolemia

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Abstract

As CABG operation is indicated in children with familial hypercholesterolemia, it seems that bilateral internal thoracic arteries are the preferred grafts, taking into consideration its superiority over venous graft regarding patency and growth.

KEYWORDS

arterial atherosclerosis, coronary artery bypass grafting operation, familial hypercholesterolemia, myocardial ischemia

1 | INTRODUCTION

Familial hypercholesterolemia (FH) is an inherited autosomal dominant disorder characterized by high serum LDL cholesterol levels that predispose the patient to premature arterial atherosclerosis, coronary artery disease, peripheral vascular disease, cerebrovascular disease, and aortic valve stenosis. Coronary artery bypass grafting operation is indicated in these patients to relieve symptoms of myocardial ischemia and to decrease the morbidity and mortality rates. We present the case of a 12-year-old girl with familial hypercholesterolemia who underwent coronary artery bypass grafting operation by using bilateral internal thoracic arteries and a saphenous vein graft. The girl convalescent uneventfully, and on 6-month follow-up, she is asymptomatic with normal physical activity.

Familial hypercholesterolemia is an inherited autosomal dominant disorder characterized by a mutation in the low-density lipoprotein (LDL) receptor gene. In heterozygous pattern, the patient inherits only one of the abnormal genes, whereas in the homozygous pattern, the patient has abnormal gene from both parents. These mutations lead to failure of the hepatocyte in clearing LDL cholesterol (LDL-C) from blood circulation and consequently a longer LDL-C half-life.[1] High LDL-C levels cause vascular endothelium dysfunction and are considered one of the most important risk factors for the development of arterial atherosclerosis.[2] Patients with FH are at risk of developing premature coronary artery disease (CAD; in the second decade of life), peripheral vascular disease, cerebrovascular disease, and aortic valve stenosis. Moreover, cutaneous and tendon xanthomas are frequent.[3-5] Coronary artery bypass grafting (CABG) operation is indicated in patients with FH to relieve symptoms of myocardial ischemia and to decrease the morbidity and mortality rates.[2,6,7] Herein, we present the case of a 12-year-old girl with FH who underwent CABG by using bilateral internal thoracic arteries (BITA) and a saphenous vein graft (SVG).

2 | CASE PRESENTATION

A 12-year-old girl was admitted to our center for diagnostic coronary angiography. She complained from typical chest pain since 2 months ago with recent worsening of symptoms causing limitation in physical activity. The patient and one of her three sisters were diagnosed with FH 7 years ago, while her only brother was not. She received medical therapy with 20 mg atorvastatin, 81 mg aspirin, and 75 mg clopidogrel. The patients' height and body weight were within normal range (143 cm, 38 kg, respectively). Physical examination revealed xanthomas over the extensor surfaces of the elbows and knees and on her feet. A 3/6 systolic murmur radiating to the neck was heard at the base of the heart. Electrocardiography (ECG) showed ST segment depression and T-wave inversion in leads V1-V6 along with signs of left ventricular hypertrophy. The transthoracic echocardiography (TTE) showed mild-to-moderate aortic valve stenosis (peak gradient across the valve was 29 mm Hg) with mild aortic regurgitation and mild mitral regurgitation. Left ventricle function was normal (ejection fraction [EF] was 60%). Doppler study of the neck vessels showed trivial narrowing in the carotids. Total cholesterol, LDL cholesterol, HDL cholesterol, and triglycerides (TG) levels were 641 mg/dL, 421 mg/dL, 191 mg/dL, and 212 mg/dL, respectively. Coronary angiography was performed, which revealed 90% ostial left main (LM) stenosis, 90% stenosis in the first segment of the left anterior descending artery (LAD), 90% stenosis in the first segment of the circumflex artery (CX), and 95% ostial stenosis in the right coronary artery (RCA; Figures 1 and 2). The patient was scheduled for urgent CABG operation. The operation was performed through median sternotomy. BITA were harvested and both have excellent flow, and a SVG was also prepared. After heparinization, the patient was placed on cardiopulmonary bypass, and the heart was arrested by antegrade cold blood cardioplegic solution. At first, the obtuse marginal artery (OM) was grafted by the SVG, and a second dose of cardioplegia was administered through it. The ascending aorta (which was markedly thickened) was opened to inspect the aortic valve. The aortic valve was trileaflet, however, with leaflet thickening and apparent lipid deposits. There was not any commissural fusion, and no stenosis was noticed. The decision was taken to preserve the native valve, and the aortotomy was closed appropriately. The RCA and LAD were bypassed using in situ right and left internal thoracic arteries, respectively. Then, the venous graft was implanted on the ascending aorta. The aortic cross clamp was released, and the patient was weaned off the bypass uneventfully without any inotropes. She was extubated after 4 h in the intensive care unit where she stayed for only 24 h. Postoperative TTE showed normal LV function with EF =60% and mild aortic valve stenosis and mild regurgitation. She was discharged from the hospital after 5 days on the following medications:

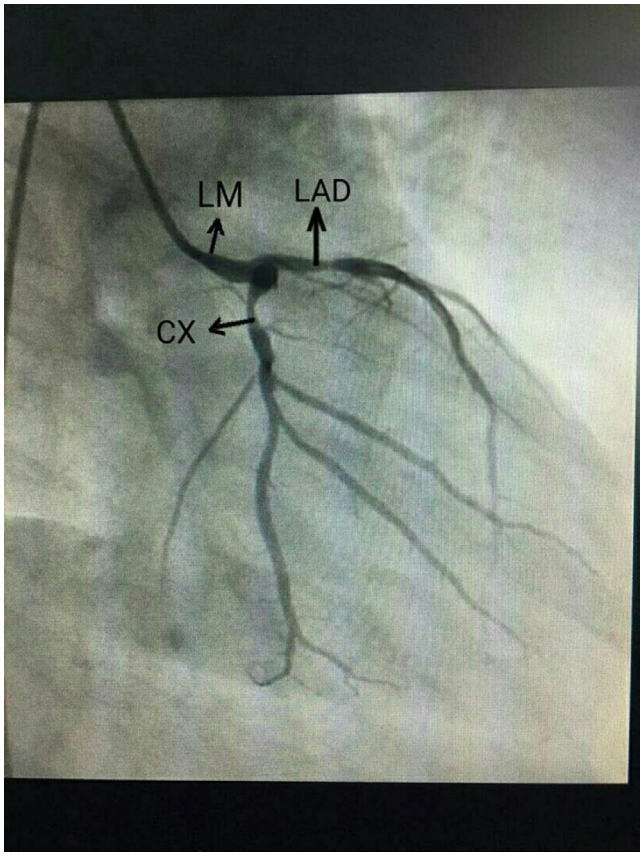

FIGURE 1 Coronary angiography showing the lesions of the left main coronary artery and its branches. CX, circumflex artery; LAD, left anterior descending artery; LM, left main artery

20 mg atorvastatin, 81 mg aspirin, 75 mg clopidogrel, and 2.5 mg bisoprolol. On 6-month follow-up, the patient was asymptomatic with normal physical activity and continued on the prescribed medications.

3 | DISCUSSION

Familial hypercholesterolemia is an inherited disorder characterized by high plasma levels of total cholesterol and LDL-C. Generally, the patients present with tendinous xanthomata and CAD early in life, especially in those who have the homozygous pattern of the disease.[1,3,8] Most of the children with the homozygous pattern will require CABG operation in the second decade of life.[4,5] In our case, we used BITA grafts to bypass the RCA and LAD arteries and a saphenous vein graft to bypass the OM artery. The superiority of arterial over venous graft on the long-term basis in both adults and children is well established, and literature review indicates that the use of left or both internal thoracic arteries is mandatory in children.[9-11] Moreover, the outcomes of BITA grafts have been shown to be better than those of conventional CABG operations in which one internal thoracic artery graft was used.[12] Good long-term results of saphenous vein grafts in

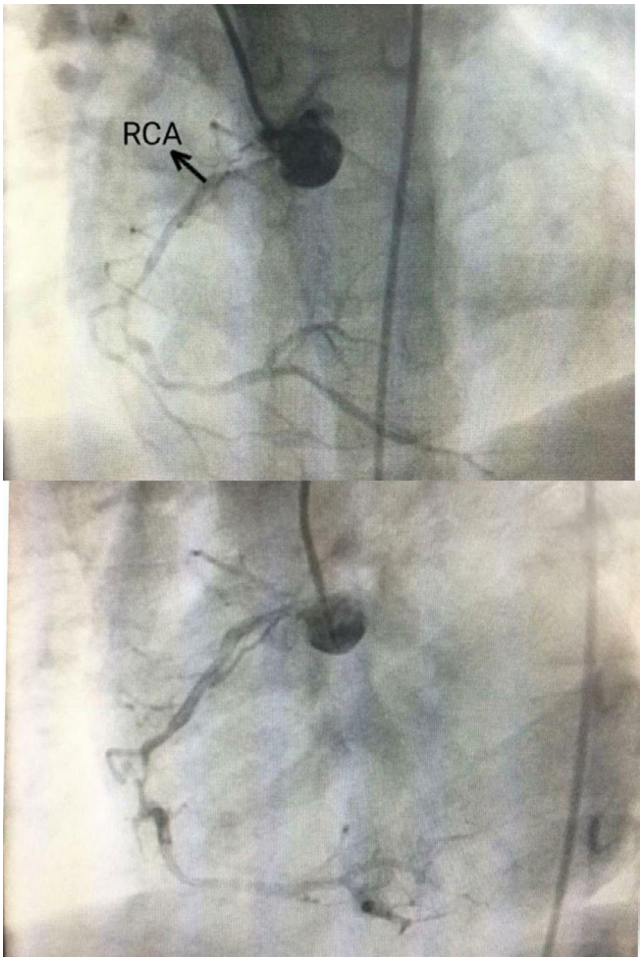

FIGURE 2 Coronary angiography showing the lesions of the right coronary artery. RCA, right coronary artery

children patients have been reported regarding patency and growth; however, it is better to avoid venous grafts in children whenever it is possible.[13] SVG when anastomosed to thickened aortic wall may be associated with poor long-term patency; however, anastomosing SVG to the internal thoracic artery is to be condemned. We did not use the radial artery since its importance as a third arterial graft is controversial. Patients with serum cholesterol levels more than 220 mg/dL are at high risk of recurrent CAD.[14] High-dose lipid-lowering medications and plasmapheresis are indicated in such patients in order to reduce the chances of recurrence. Successful staged treatment by CABG followed by liver transplantation has been described.[9,15] Recently, novel inhibitor of proprotein convertase subtilisin kexin type 9 (PCSK9) was introduced for the treatment of dyslipidemia and atherosclerosis. PCSK9 promotes the degradation of LDL receptors, and thus, PCSK9 inhibition increases LDL receptor recycling and LDL and reduces LDL-C dramatically in FH patients who could not reach targets and probably would provide better future.[16] Intraoperatively, the aortic valve area was not reduced in spite of leaflet thickening and apparent lipid deposits, and therefore, we preserved the native aortic valve. We found that aortic valve replacement was not necessary in such patient especially when taking the patient's young age into consideration.

4 | CONCLUSION

As CABG operation is indicated in children with familial hypercholesterolemia, it seems that bilateral internal thoracic arteries are the preferred grafts in children patients taking into consideration its superiority over venous graft regarding patency and growth.

ACKNOWLEDGMENT
None.

CONFLICT OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
AA-D: planned and performed the work leading to the report and wrote and reviewed successive versions and participated in their revisions. SS: participated in writing the report and approved the final version. HH, OA and MKA: wrote and reviewed the successive versions and participated in their revisions.

ETHICAL APPROVAL
The manuscript was approved by ethics committee at Damascus University.

AUTHOR'S STATEMENT
Consent for publication was granted by the patient's mother.

DATA AVAILABILITY STATEMENT
The data that support the findings of this study are available from the corresponding author, [A.A], upon reasonable request.

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REFERENCES
1. Pejic RN. Familial hypercholesterolemia. *Ochsner J*. 2014;14(4):669-672.
2. Kawasuji M, Sakakibara N, Takemura H, Matsumoto Y, Mabuchi H, Watanabe Y. Coronary artery bypass hypercholesterolemia grafting in familial hypercholesterolemia. *J Thorac Cardiovasc Surg*. 1995;109(2):364-369.
3. Nemati MH, Astaneh B, Joubeh A. Triple coronary artery bypass graft in a 10-year-old child with familial hypercholesterolemia. *Gen Thorac Cardiovasc Surg*. 2009;57:94-97.

4. Nemati MH. Coronary revascularization in a child with homozygous familial hypercholesterolemia. *Interact Cardiovasc Thorac Surg*. 2009;10(1):131-132.
5. Mabuchi H, Koizumi J, Shimizu M, Takeda R, Hokuriku FH, CHD study group. Development of coronary heart disease in familial hypercholesterolemia. *Circulation*. 1989;79:225-232.
6. Allen TM, Thompson GR, Myant NB, Sterner R, Oakley CM. Cardiovascular complications of homozygous familial hypercholesterolemia. *Br Heart J*. 1980;44:361-368.
7. Kawasuji M, Kawajiri F, Watanabe G, Matsunaga Y, Tedoriya T, Iwa T. Coronary bypass surgery for familial hypercholesterolemia. *Jpn J Thorac Surg*. 1988;41:537-540.
8. Austin MA, Hutter CM, Zimmern RL, Humphries SE Genetic causes of monogenic heterozygous familial hypercholesterolemia: a huge prevalence review. *Am J Epidemiol*. 2004;160(5):407-420.
9. Komarov R, Ismailbaev A, Chragyan V, et al. State-of-the-art pediatric coronary artery bypass surgery: a literature review. *Braz J Cardiovasc Surg*. 2020;35(4):539-548.
10. El-Khouri HM, Danilowicz DA, 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;65:1127-1130.
11. Ersoy U, Güvener M. Coronary revascularization in seven-year-old boy with homozygous familial hypercholesterolaemia. *Acta Paediatr*. 2000;89:1501-1502.
12. Nambiar P, Kumar S, Mittal CM, Sarkar IC. Outcomes of bilateral internal thoracic arteries in minimally invasive coronary artery bypass grafting with analogy to the SYNTAX trial 201. *Innovations*. 2019;14(3):227-235.
13. Bhan A, Swain S, Juneja R, Saxena P, Venugopal P. Total arterial revascularization in a child with familial homozygous hypercholesterolemia. *J Thorac Cardiovasc Surg*. 2005;129:932-934.
14. Takahashi T, Nakano S, Shimazaki Y, et al. Long-term appraisal of coronary bypass operations in familial hypercholesterolemia. *Ann Thorac Surg*. 1993;56:499-505.
15. Arora G, Fraser CD, Kearney DL, Vincent JA. Severe supravalvar aortic stenosis in familial homozygous hypercholesterolemia. *Pediatr Cardiol*. 2006;27(2):282-285.
16. Ogura M. PCSK9 inhibition in the management of familial hypercholesterolemia. *J Cardiol*. 2018;71(1):1-7.

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