

# Pubertede Koroner Revaskularizasyon ve Ailesel Hiperlipidemi: Greft Seçimi

## FAMILIAL HYPERLIPIDEMIA AND PUBERTAL CORONARY REVASCULARIZATION: GRAFT SELECTION

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### Özet

Familial hiperlipidemi, çok erken yaşlarda koroner arter hastalığına sebep olabilen malign karakterli nadir bir hastalıktır. Burada plazma kolesterol düzeyleri kontrol altına alınmamış 14 yaşında bir olguyu sunuyoruz. Büyüme potansiyeli ve açık kalma oranları göz önüne alındığında greft seçiminin bu olguda önem gösterdiği ortaya çıkmaktadır. Biz olgumuzda iki adet arteriyel, bir adet de safen ven grefti kullandık.

**Anahtar kelimeler:** Familial hiperlipidemi, koroner arter hastalığı, koroner bypass, greft

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### Summary

Familial hyperlipidemia is a rare condition with malignant behavior, resulting in premature coronary artery disease. We report a 14 years old case with undertreated plasma cholesterol levels. Graft selection due to growth potential and patency rates was one of the important problems in this patient. Two arterial grafts and one saphenous vein were implanted for coronary revascularization.

**Keywords:** Familial hyperlipidemia, coronary artery disease, coronary bypass, graft

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### Introduction

Homozygous familial hypercholesterolemia (FH) is a rare condition with an incidence of 1/1 million births. Excessive levels of plasma cholesterol (> 600 mg/dL) and cutaneous xanthomata are characteristic before the age of 5 years [1]. We operated a 14 years old girl with FH. Although this one is not a unique case, we report it to call attention of the community to graft selection in pediatric cases.

### Case

A 14 years old girl was operated due to ostial occlusions of left main trunk and right coronary artery. In her history, she was hospitalized for evaluation of tumor-like cutaneous plaques over distal joints and diagnosed as xanthoma 9 years ago. Cutaneous symptoms began over her coxycs and spread out to extremities (Figure 1). She had high plasma lipid levels (cholesterol 772 mg/dL; triglycerides 74 mg/dL; HDL 14 mg/dl; LDL 571 mg/dL) and abnormal lipid electrophoresis (a 4.5%, pre-b 9.9%, b 85.4%). Other biochemical parameters were normal. She was weighing 16 kg (3-10 percentile) and 103 cm high (3-10 percentile). The diagnosis was "familial hyperlipidemia type 2a". Plasma lipid content of her brothers and father were within normal limits. Her mother's lipid profile was unknown, because she had died accidentally 3 years ago. A dietary program excluding saturated fatty acids and medical treatment was

planned. No neurological, cardiac or ophthalmic pathology was noted. After medical treatment and dietary suggestions, the result was desperate after 5 months: cholesterol 614 mg/dL and triglyceride 123 mg/dL. Probably, she was not obeying to the regimen. Closer follow-up with monthly intervals was planned. Because the family was living in a rural area, it was not easy to perform frequent visits. Cholesterol levels ranged between 444 to 817 mg/dL despite all medical attempts. Plasmapheresis was not performed due to discordance of her family. She came to control for the last time on July 1992. She reappeared with stable angina pectoris on August 1998. Physical examination was almost normal. Cutaneous lesions were persevering and a slight systolic murmur over pulmonary valve was heard. Morphological findings on echocardiography were normal, with 70% ejection fraction and 39% fractional shortening. Cholesterol level was 583 mg/dL (LDL 535 mg/dL, HDL 35 mg/dL) and triglyceride 64 mg/dL. A treadmill test was planned and pravastatin 10 mg/day was added to regimen. After another long period of disappearance, treadmill test was performed on May 2000, with a non-surprising positive result. Coronary angiography showed a 40% narrowing of left main trunk, and cholesterol levels were very high (total 629 mg/dL, HDL 46 mg/dL, LDL 569 mg/dL, triglyceride 70 mg/dL). Pravastatin was doubled (20 mg/day), aspirin 300 mg/day and metoprolol was added. There were no cardiac symptoms until November 2001. Then she was hospitalized with severe unstable angina pectoris. Angiographic examination showed 70% occlusions of



**Figure 1 A and B.** Cutaneous xanthomata on the extremities.

both left main trunk and right coronary artery, and a normal ventriculography. An urgent coronary artery bypass grafting was mandatory for this child. Graft selection was right internal mammary artery (IMA) for the right coronary artery, left IMA for the left anterior descending artery and greater saphenous vein graft for the circumflex artery. She weaned from cardiopulmonary bypass with the aid of intra-aortic balloon counterpulsation (IABP), which was removed 4 hours postoperatively. Inotropic support was ceased on the first postoperative day.

This patient is now under strict follow-up and two sessions of plasmapheresis have been performed. Plasma cholesterol levels lowered significantly (total cholesterol 291 mg/dL; LDL 231 mg/dL; HDL 37 mg/dL) and the result of a control treadmill test 3 months after the operation was negative.

## Discussion

As coronary artery disease (CAD) is the major cause of death in western world, dyslipidemia is one of the most important risk factors of it [2]. Particularly younger patients with dyslipidemia may have rapidly occlusive disease. Most of these cases are resistant to anti-hypercholesterolemic treatment. The proportion of plasma cholesterol contents influences the behavior of the disease and strongly correlated with clinical outcome of patients. There patients developing CAD have significantly lower values of plasma high density lipoprotein cholesterol levels and a higher ratio of total cholesterol/HDL when compared with patients free of CAD [3]. Usually, medical treatment doesn't give any satisfying results. Familial hyperlipidemia may have a morbid nature. Significant narrowing of the coronary arteries may result in premature cardiac death during childhood. Clinical condition is very similar to that of middle age individuals' atherosclerotic coronary artery disease. But pathological

examinations have showed that the vascular lesion is not an atheromatous plaque, but a xanthomatous deposit of excessive plasma cholesterol [4]. Coronary revascularization improves the long term survival in those patients. From the surgical point of view, selection of grafts is the main question. Arterial revascularization offers better late-cardiac-event-free survival. But according to the literary data, we have avoided from extensive use of arterial conduits (radial or right gastroepiploic arteries), which does not have additional benefit or cost [5]. We have anastomosed the right internal thoracic artery to the right coronary artery, and the left internal thoracic artery to the left anterior descending coronary artery. Blood supply to circumflex system was maintained with a greater saphenous vein graft. A relatively longer saphenous vein was anastomosed to circumflex artery to overcome the potential shrinkage of the graft with growing up of the child. Complete occlusion of venous grafts is almost a rule in patients under 5 years of age, but patency rate improves in those over 10 years. Rapid degeneration of the vein and increasing body size of the patient may be reasons of graft failure [7]. On the other side, arterial grafts have the growth potential. We have explained early postoperative period of low cardiac out-put by chronic ischemia of the *young* myocardium. The stunning heart responded very well after 4 hours of IABP support.

This case was the first pediatric coronary bypass surgery due to familial hyperlipidemia in our institution. Good information of the family members and peer follow-up will be mandatory for this patient.

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