





## A dramatic example of severe premature atherosclerosis successfully treated by percutaneous coronary intervention 🎥

 Gökhan Altunbaş,  Ertan Vuruşkan,  Osman Başpınar\*,  
 Murat Sucu  
Departments of Cardiology, and \*Pediatrics, Faculty of Medicine, Gaziantep University; Gaziantep-Turkey

### Introduction

Familial hypercholesterolemia (FH) is one of the most frequent causes of premature atherosclerosis. We present a case of an 11-year-old boy with acute coronary syndrome presenting with cardiac arrest and diffuse coronary atherosclerosis, who was treated successfully by percutaneous coronary intervention (PCI).



**Figure 1.** Arcus cornea is clearly visible

### Case Report

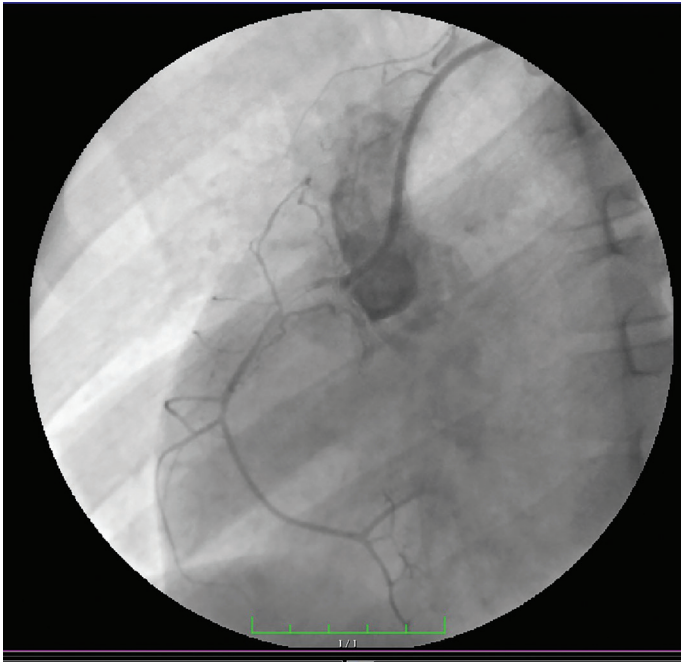
An 11-year-old boy was admitted to the emergency department with complaints of chest discomfort and dyspnea. Electrocardiography showed deep symmetric T-wave inversions through V1-V4. Physical examination was unremarkable except for arcus cornea (Fig. 1) and xanthomas on the knee joint (Fig. 2). Table 1 shows the baseline characteristics of the patient. Emergent cor-

**Table 1. Baseline characteristics of the patient**

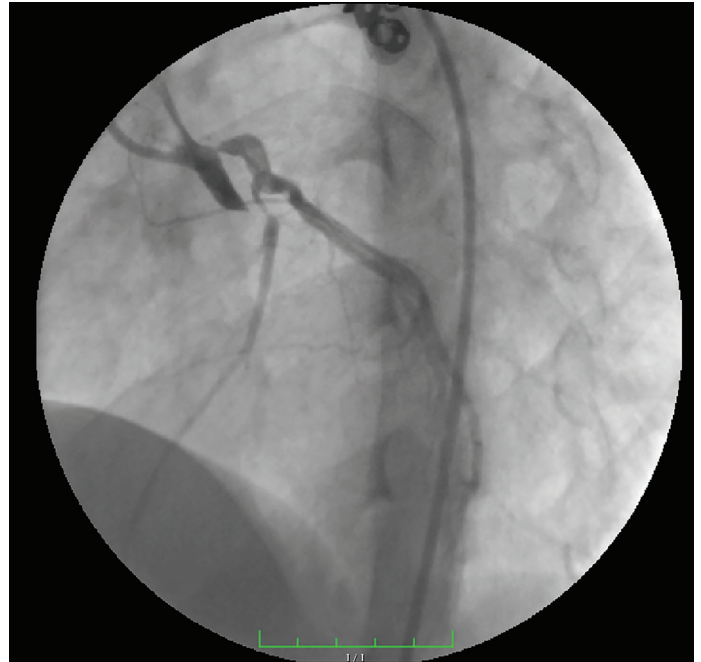
Age (years)	11
Sex	Male
Diabetes	No
Hypertension	No
Medications	None
Smoking	No
Baseline fasting plasma lipid levels (mg/dL)	Total cholesterol: 493 HDL-C: 62 LDL-C: 378 Triglyceride: 81
HDL-C - high density lipoprotein-cholesterol; LDL-C – low density lipoprotein-cholesterol	



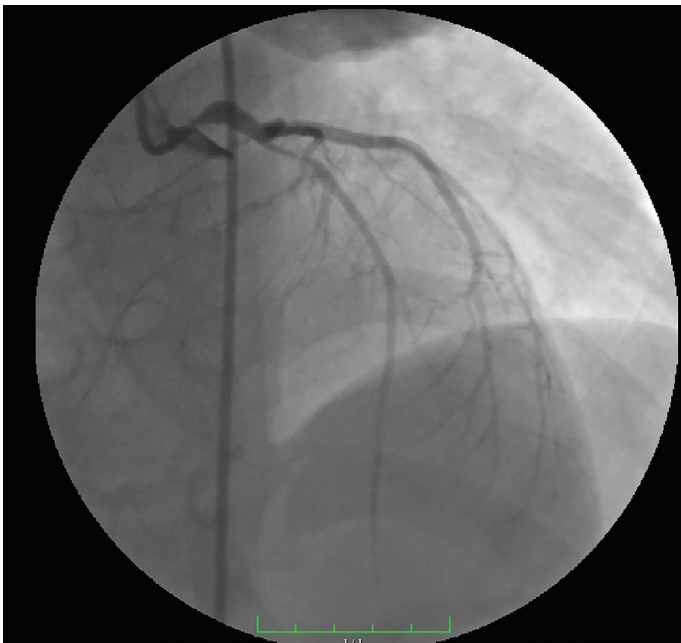
**Figure 2.** Tendon xanthoma is present



**Figure 3.** Right coronary artery proximal critical stenosis is shown

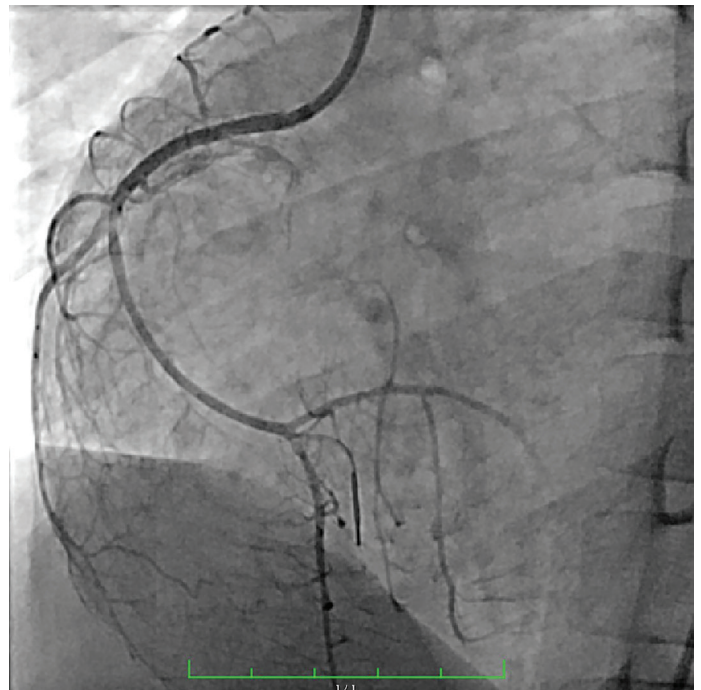


**Figure 5.** LMCA ostial and LAD proximal critical stenoses are shown



**Figure 4.** LMCA ostial and LAD proximal stenoses are shown

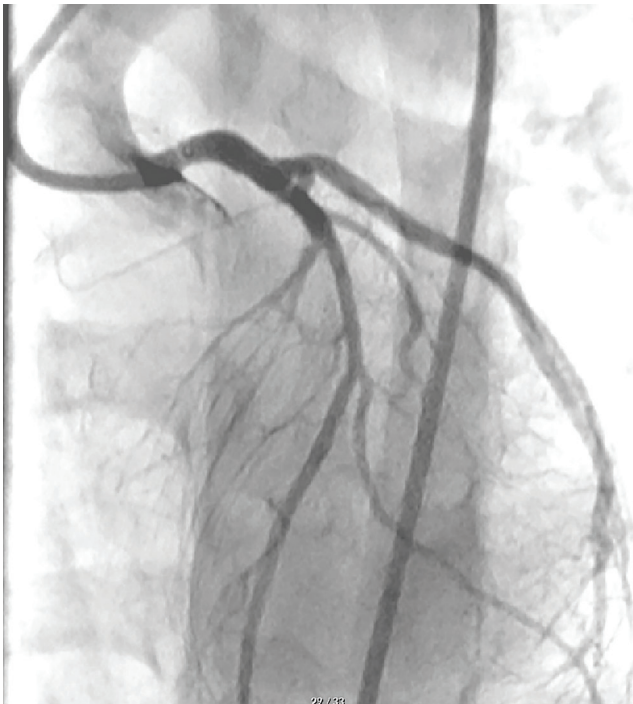
Coronary angiography was performed and revealed diffuse atherosclerosis including severe narrowing of the right coronary artery (RCA) proximal segment, left main coronary artery (LMCA), and left anterior descending artery (LAD) proximal segment (Fig. 3-5). Coronary artery bypass graft (CABG) surgery was first considered, and the patient consulted with the cardiac surgery team. Owing to his younger age, surgeons considered the surgery as prohibitively high risk, and PCI was planned. According to pediatric doses based on the age and weight of the patient; 80 mg aspirin and half a tablet of 75 mg clopidogrel were administered.



**Figure 6.** Right coronary artery after successful stenting

Weight-adjusted bolus of unfractionated heparin was administered, and then RCA was cannulated using a 5 French (F) IMA guiding catheter. A floppy guidewire (Asahi Soft, Asahi, Japan) was used to cross the lesion. The lesion was first dilated with 1.5×20 and 2.0×20 percutaneous transluminal angioplasty balloons, and 2.25×28 mm Xience stent (Abbott, USA) was implanted. A 2.5×15 non-compliant (NC) balloon was used to post-dilate the stent at high pressure to achieve complete stent apposition (Fig. 6). Owing to the younger





**Figure 7.** LMCA and LAD after successful stenting

age of the patient, the angle of the arcus aorta was narrow, and it was hard to cannulate LMCA. After various catheters, finally, we were able to cannulate LMCA using a 6F hockey stick guiding catheter. We again crossed LAD using a floppy guidewire (Asahi Soft). LAD proximal lesion was first predilated with a 2.5×15mm balloon, and then a 2.75×18 Xience stent (Abbott) was implanted. LMCA lesion was first dilated with a 3.0×12 balloon, and then a 3.0×16 Promus stent (Boston Scientific, USA) was implanted from LMCA ostium to LAD. A 3.5×12 NC balloon was used to post-dilate LMCA stent (Fig. 7). Intravascular ultrasound (IVUS) showed complete stent apposition (Video 1). The patient was then transferred to the intensive care unit. Postoperative recovery was uneventful.

## Discussion

In this case report, we present a dramatic example of diffuse coronary atherosclerosis at 11 years old and successful treatment by PCI.

There are three different formal criteria to diagnose FH; however, the Dutch Lipid Clinic Network criteria are the most widely accepted criteria (1). The estimated prevalence of homozygous FH in the general population is 1:1.000.000; however, it is believed that the prevalence is severely underestimated, and the disease is undertreated in the general population (2). Based on the Dutch Lipid Clinic Network, patients with  $\geq 8$  points are accepted as definite FH. Our patient has 21 points (family history: two of the patient's brothers were on lipid apheresis due to FH, 1 point; clinical history: 2 points; physical examination: tendinous

xanthoma+arcus cornea, 10 points; cholesterol levels: 8 points; total: 21 points) and thus accepted as definite FH.

Treatment with statins is safe and effective in children aged  $\geq 8$  years, and treatment goals are at least 50% reduction or low-density lipoprotein-cholesterol (LDL-C)  $< 130$  mg/dL (3). For patients with homozygous FH, intensive statin treatment is usually ineffective, and thus LDL apheresis will be required in the majority of the patients. In our country, there are two large-scale registries evaluating FH, A-HIT1 and A-HIT2. A-HIT1 evaluated patients with homozygous FH, and A-HIT2 enrolled patients with FH (homozygous and heterozygous). The major finding was that patients with FH are undertreated, and many of them did not reach target LDL-C levels despite lipid apheresis (4).

For our patient, intensive statin treatment and LDL apheresis were immediately started. The major dilemma for our patient was to choose between percutaneous treatment or CABG surgery. CABG surgery has long been used in the pediatric population mostly due to congenital anomalies, Kawasaki disease, or as a rescue operation for complications after pediatric heart operations. Most of the experience originates from patients from patients with Kawasaki disease, which is the most frequent indication for pediatric CABG surgery (5). Even for children  $\leq 3$  years old, CABG surgery appears to be safe and has good long-term patency rates (6).

PCI has been mainly used in children with Kawasaki disease or for the treatment of complications after congenital heart disease surgery. Schneider et al. (7) reported five patients  $\leq 18$  years old undergoing PCI in an 8-year duration. In their series, only one patient required PCI for the treatment of acute myocardial infarction due to atherosclerotic narrowing of RCA and LAD. Jalal et al. (8) reported periprocedural and late outcomes of pediatric patients undergoing PCI in two institutions in a 17-year time range. They revealed 40 PCIs performed in 29 patients. The mean age of the treated population was 6.5 years. In this series, the most common indication for PCI was postoperative complications in patients with transposition of great arteries.

As regards to the features of PCI technique in the pediatric population, there are no major differences. Two important points to consider are selecting the appropriate guiding catheter and determining the size of coronary balloons and stents based on not only visual estimates but also intracoronary imaging techniques (IVUS or optical coherence tomography). In our case, we were able to cannulate RCA using a 5F IMA guiding catheter. After testing various size and angled catheters, we were able to cannulate LMCA only using a 6F hockey stick guiding catheter. The sizes of the balloons and stents were determined by IVUS and post-implantation stent appositions were also evaluated by IVUS.

Percutaneous treatment of atherosclerotic coronary artery disease in the pediatric population with FH is found in the literature only as case reports. In such a case report, Nazif et al. (9) reported the successful treatment of a 3-year-old patient with PCI using a bioresorbable scaffold (BRS). Their patient had LMCA and RCA ostial critical stenosis. They placed a BRS into both coronary arteries with guidance of IVUS. This patient resembled quite

similar features with our patient. They chose BRS over metallic stents mainly due to the concerns of the growth of the child that will accompany enlargement of the coronary arteries, and thus metallic stents will become undersized. Indeed, Oberhoffer et al. (10) demonstrated that both right and left coronary arteries grow up to five times in diameter from infancy to teenager. For our patient, we also considered BRS. However, owing to both recent reports of increased stent thrombosis with BRSs and health insurance-reimbursement problems, we selected metallic stents.

## Conclusion

Percutaneous coronary intervention is safe and effective for the treatment of pediatric patients with atherosclerotic coronary artery disease.

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**Video 1.** IVUS showing complete stent apposition for both LAD and LMCA stents.

**Address for Correspondence:** Dr. Gökhan Altunbaş,  
Gaziantep Üniversitesi Tıp Fakültesi,  
Kardiyoloji Anabilim Dalı,  
Gaziantep-Türkiye  
Phone: +90 342 360 60 60  
E-mail: drgokhanaltun@gmail.com

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