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

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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	Total cholesterol: 493
lipid levels (mg/dL)	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

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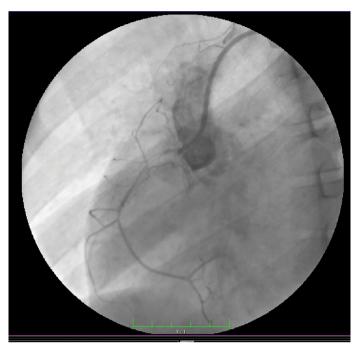


Figure 3. Right coronary artery proximal ciritical stenosis is shown



Figure 4. LMCA osteal and LAD proximal stenoses are shown

onary 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.

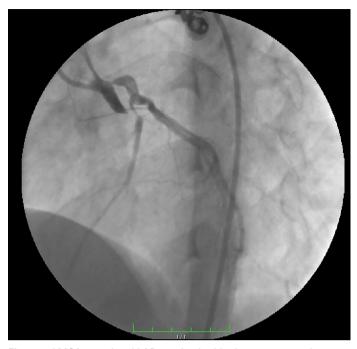


Figure 5. LMCA osteal and LAD proximal critical stenoses are shown

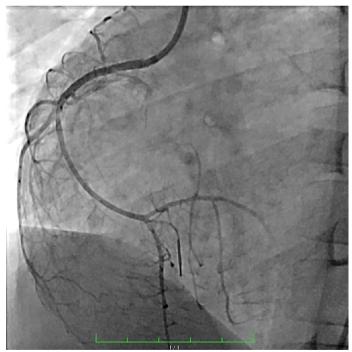


Figure 6. Right coronary artery after successful stenting

Weight-adjusted bolus of unfractioned 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

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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 ≥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 ≥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 ≤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 osteal critical stenosis. They placed a BRS into both coronary arteries with guidance of IVUS. This patient resembled quite

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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.

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A Port-A-Cath silent embolization to the left distal pulmonary artery:

A novel percutaneous approach for a challenging case

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Introduction

Totally implantable venous access port devices have been commonly used to monitor hemodynamic parameters and to infuse medications, blood, other blood products, and fluids. They are also crucial for the chronic and acute care of patients with many diseases, namely for patients with cancer in need of longterm chemotherapy treatment. Nevertheless, several complications are associated with the use of these devices. One of these complications is catheter migration, most commonly to the pulmonary artery (PA), the right ventricle, and the right atrium (1, 2). One exceptional yet potentially severe complication in using this device for treatment and port catheter placement is the embolization of one of the parts of the device. However, since the majority of them are asymptomatic, the condition is usually not detected for a long time and is usually incidentally diagnosed. The embolization may lead to dangerous complications in the heart and lungs, such as cardiac arrhythmia, myocardial disorders, arterial rupture (in the heart or the lungs), thrombosis, perforations in the heart valves, pulmonary embolism, and endocardial infection. The initial intervention usually includes a