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CASE REPORT

ADVANCED

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

OCT

A Modality for Identifying Stent Failure in Pediatric Patients With Angiographically Silent Coronary Arteries

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ABSTRACT

We present the case of a 16-year-old patient with anomalous left coronary artery from the left pulmonary artery requiring percutaneous coronary intervention in infancy who presented with ventricular fibrillation arrest. A coronary angiogram revealed 40% narrowing of the stent relative to the remainder of the left main coronary artery. Optical coherence tomography was performed and revealed an area stenosis of 70% relative to the native left main coronary artery. The patient had outgrown the stent. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:849–52)

HISTORY OF PRESENTATION

A 7-month-old infant had a diagnosis of anomalous left coronary artery from the left pulmonary artery (ALCAPA). A transthoracic echocardiogram (TTE) revealed that the patient had a dilated left ventricle with apical and lateral wall motion abnormality, mild to moderate mitral regurgitation, and echogenic papillary muscles suggesting an ischemic injury. After the diagnosis was made, the patient underwent a coronary reimplantation procedure. Following the procedure, the patient deteriorated clinically and required extracorporeal membrane oxygenation. Evaluation with coronary angiography revealed stenosis of a stretched segment of the left main coronary artery (LMCA). The LMCA was implanted to the desired location of the aorta; however, there was insufficient length of LMCA for the relocation, and this resulted in the stretched segment of the LMCA. At the time, reperforming the operation was considered; however, the stenotic lesion did not respond to conventional balloon angioplasty. Therefore, coronary stenting was pursued. The stenosis was successfully corrected with the placement of a sirolimus-coated 2.25 × 8 mm Cypher endovascular coronary artery drug-eluting stent (DES) (Cordis, Santa Clara, California). A DES was used preferentially over a bare-

LEARNING OBJECTIVES

- To identify outgrowing coronary stent as a cause off stent failure in pediatric patients.
- To recognize that OCT may provide utility as an adjunct to conventional cardiac imaging for surveillance of coronary stents in pediatric patients.

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ABBREVIATIONS AND ACRONYMS

ACE = angiotensin-converting enzyme

ALCAPA = with anomalous left coronary artery from the left pulmonary artery

CMR = cardiac magnetic resonance

DES = drug-eluting stent

ECG = electrocardiogram

LMCA = left main coronary artery

LV = left ventricular

MLA = minimum lumen area

OCT = optical coherence tomography

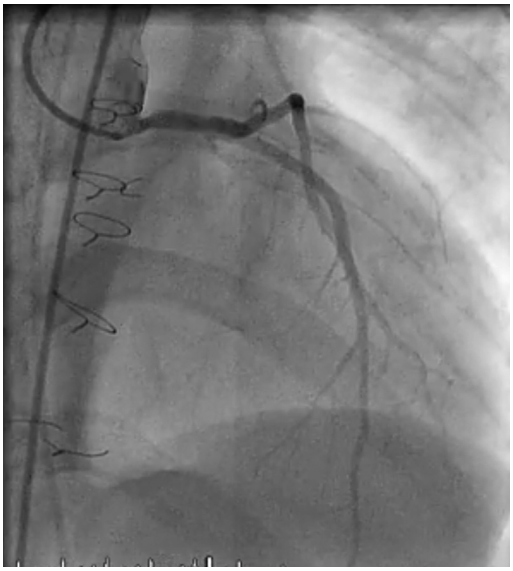
PCI = percutaneous coronary intervention

TTE = transthoracic echocardiography

metal stent with the hope of reducing in-stent stenosis. However, as a result of the stretched LMCA, the patient sustained myocardial injury. A follow-up TTE demonstrated a distended left ventricle with reduced contractility, a dilated left atrium, and severe mitral regurgitation. The patient was administered an angiotensin-converting enzyme (ACE) inhibitor, aspirin, and clopidogrel for 6 months, followed by long-term therapy with an ACE inhibitor and aspirin.

Given the unknown long-term outcomes of coronary stents in pediatric patients, our patient had annual surveillance with invasive and noninvasive cardiac imaging modalities. Initially, the patient was followed with annual electrocardiograms (ECGs), TTEs, and coronary angiograms. Overtime, because of clinical stability, cardiac magnetic resonance (CMR) stress testing was used as the primary method to assess the status of the coronary stent, and invasive testing with coronary angiograms was changed to every 2 to 3 years. During annual follow-up visits, the patient was free of any cardiac symptoms. The ECG showed normal sinus rhythm, left-axis deviation, and no new ischemic changes. The stent was well visualized on the TTE with good antegrade flow. A left ventricular (LV) angiogram showed improvement in LV function. On coronary angiography, there were no signs of any significant renarrowing of the stent, and the coronary vessel distal appeared to have grown without any obstruction. At 15 years of age, cardiac catheterization showed <40% narrowing of the coronary stent, unchanged from the diameter since implantation in infancy. Results of CMR stress tests were also stable over time, showing mild LV enlargement, with mild systolic dysfunction with an ejection fraction of 53%. A decision was made to delay reintervention, to avoid the complications associated with stent re-expansion (coronary perforation, edge dissections), in an asymptomatic patient. However, at 16 years of age, he had an out-of-hospital ventricular fibrillation arrest while exercising in gym class. He underwent successful resuscitation on the scene and was transferred to a nearby tertiary care center.

FIGURE 1 **Coronary Angiogram**



Coronary angiogram showing the stented left main coronary artery after the patient was stabilized from ventricular fibrillation arrest.

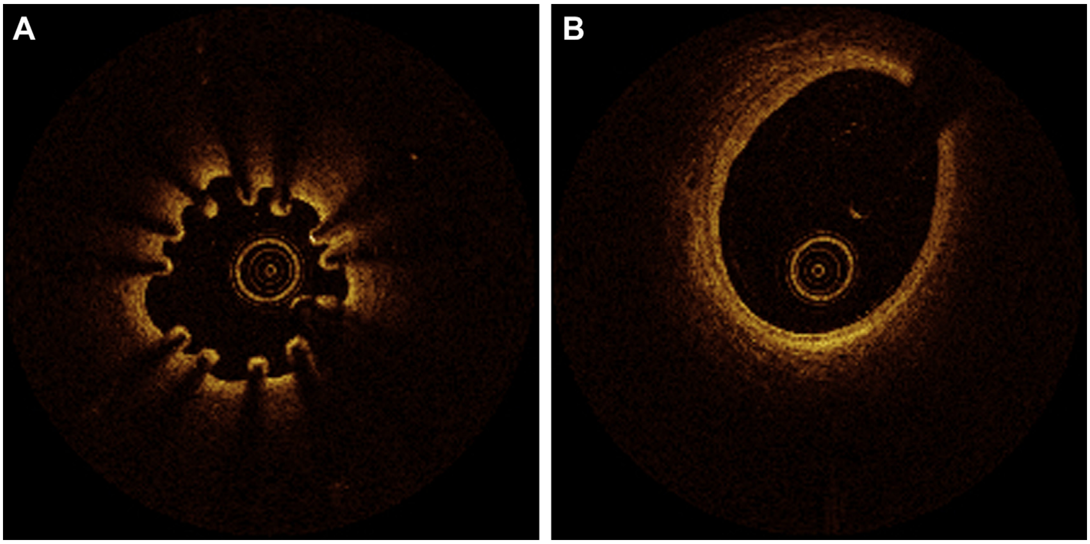
QUESTION 1: WHAT IS THE DIFFERENTIAL DIAGNOSIS FOR THIS PATIENT'S CARDIAC ARREST?

The differential diagnosis included acute ischemia resulting from stent failure versus scar tissue-mediated ventricular tachyarrhythmia. One of the main concerns following PCI in pediatric patients is that the child may outgrow the stent.

QUESTION 2: WHAT IS THE NEXT APPROPRIATE STEP TO INVESTIGATE THE CAUSE OF CARDIAC ARREST?

Unlike in adults PCI is rare in pediatrics, and the natural history of coronary artery DESs in pediatrics is unknown. After the patient's condition was stabilized, a decision was made to perform an angiogram to assess the patency of the stent. Coronary angiography demonstrated that the right coronary artery, left anterior descending, and left circumflex artery were all angiographically normal. There was 40% angiographic stenosis of the stented LMCA compared with the reference vessel (**Figure 1,** Videos 1 and 2). The area of stenosis was unchanged from his previous surveillance coronary angiograms. This did not plain the cause of his ventricular fibrillation arrest; therefore, OCT was performed to clarify the mechanism of his presentation. OCT revealed that the minimum lumen area (MLA) in the stented LMCA was 4.5 mm^2, with a reference vessel diameter distally of approximately 15 mm^2, thus giving an area stenosis of 70%, which would be anatomically significant (**Figures 2A and 2B,** Video 3). Moreover, the MLA on OCT was much lower than the established threshold for

FIGURE 2 Optical Coherence Tomography of the Left Main Coronary Artery



(A) The stented segment of the left main coronary artery. There is clear evidence of stent malapposition. The luminal diameter of the stent is 4.75 mm^2. **(B)** The reference vessel, which is the distal portion of the left main coronary artery. The luminal diameter of the reference vessel is 15 mm^2.

significant LMCA developed by intravascular ultrasound. There was no evidence of restenosis, neoatherosclerosis, exposed stent struts, or intraluminal thrombus, all potential mechanisms for stent failure. The mechanism was late acquired malapposition, with the patient's LMCA essentially outgrowing the stent. Fractional flow reserve was performed and confirmed physiological significance of the LMCA lesion. The patient underwent left main coronary arterioplasty using autologous saphenous vein and implantation of an automatic implantable cardiac-defibrillator, and he has remained event free. The patient is currently 18 years old and is followed by the adult congenital heart disease clinic.

QUESTION 3: HOW SHOULD CORONARY STENTS BE MONITORED FOR STENT FAILURE IN PEDIATRIC PATIENTS?

The use of PCI in pediatrics is rare. Recent case reports documented the use of PCI in infants, but only a handful of cases involved patients with ALCAPA. Two single case reports, of 6- and 7-year-old children, respectively, described positive short-term outcomes (1,2). However, the natural long-term history of coronary artery DES in pediatrics is largely unknown. Given the uncertainty of his clinical course, the patient was followed closely to assess for ischemia from stent failure. Following stent placement, the patient was monitored annually with multiple diagnostic studies, including TTEs, ECGs, coronary angiograms, and CMR stress tests, none of which demonstrated evidence of significant renarrowing of the stent or new ischemia. Initially, coronary angiography was the main imaging modality used to screen for myocardial ischemia and stent failure. Over time, however, the patient was deemed clinically stable because of the absence of symptoms, as well the unchanged degree of stenosis in the stent. To minimize invasive imaging, the main method for screening became stress CMR, which occurred annually, whereas coronary angiography was performed every 2 to 3 years. Stress CMR has been well studied in acquired heart disease (3); however, its evidence for use in congenital heart disease is limited. A small study by Deva et al. (4) assessed the role of stress CMR for detecting ischemia in congenital heart disease. The study demonstrated that the sensitivity of the test was 82%. Therefore, negative test results cannot fully rule out myocardial ischemia. Regarding coronary angiography, it is currently the gold standard for assessing the patency of coronary arteries and stents. However, conventional coronary angiography provides a 2-dimensional representation of a 3-dimensional structure, which can lead to imprecise estimation of disease severity (5). In our patient, investigation with OCT showed that the reference LMCA had significantly outgrown the stented LMCA, and this mismatch resulted in an area of stenosis of approximately 70%. OCT can characterize the morphological features of the coronary lesion, and it allows for a more accurate evaluation of disease severity (5). Intravascular imaging is recommended for evaluation of stent failure (6). This recommendation has increasingly led to using intravascular imaging techniques such as OCT to supplement the information obtained from conventional coronary angiograms. In pediatric cardiology, there has been increasing interest in the use of OCT for evaluating coronary anatomy in patients with Kawasaki disease and post-heart transplantation (7–9). Our case shows the potential for using OCT as an adjunct to conventional cardiac imaging for surveillance of coronary stents in pediatric patients.

QUESTION 4: IN CLINICALLY STABLE PEDIATRIC PATIENTS WITH CORONARY STENTS, WHEN IS THE OPTIMAL TIME FOR REINTERVENTION?

As illustrated by our case, effective regular evaluation of stent patency is important in a growing child because adjacent coronary arteries can outgrow the stented coronary artery diameter. Given the scarcity of published reports of PCI use in pediatric patients, the optimal timing for reintervention in clinically stable patients is still debated.

CONCLUSIONS

PCI in infants and children remains rare, but it has shown feasibility as a potential effective long-term treatment option for coronary stenosis in pediatric patients. Our patient underwent PCI as an infant and is currently the longest surviving case documented. Furthermore, our case highlights challenges in detecting coronary artery stent failure caused by a significantly outgrown stent in an asymptomatic patient. With growing evidence of the use of OCT in pediatric patients, consideration should be given to using OCT for detecting stent failure in patients with angiographically silent coronary arteries.

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KEY WORDS ALCAPA, long-term survival, OCT, outgrown stent, PCI

APPENDIX For supplemental videos, please see the online version of this article.