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CASE REPORT
Congenital heart disease

Intravascular ultrasound-guided stent implantation in reimplanted left main coronary artery of a 15-year-old child: a case report

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Background Only a few cases have been reported about clinical value of percutaneous coronary intervention (PCI) and intravascular ultrasound (IVUS) in patients with stenosis of a re-implanted left main coronary artery (LMCA).

Case summary We herein report a rare case of restenosis after direct reimplantation of an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) in a 15-year-old girl. At the first evaluation, she had mildly reduced systolic dysfunction with left ventricular ejection fraction of 47%. Three months after surgical repair, the patient developed recurrent precordial pain. Consequent imaging tests and IVUS revealed a restenosis of the LMCA characterized as an attenuated plaque with a large plaque burden. A drug-eluting stent was implanted with IVUS guidance. Follow-up revealed a patent LMCA and preserved systolic function.

Discussion The current case demonstrated that IVUS-guided PCI can be feasible in the treatment of coronary artery stenosis after repair of an ALCAPA. Further study is needed to explore the pathophysiological mechanism of this condition and the clinical value of PCI and IVUS in patients with stenosis of the LMCA.

Keywords Anomalous left coronary artery from pulmonary artery • Percutaneous coronary intervention • Intravascular ultrasound • Stenosis of left main coronary artery • Case report

ESC Curriculum 2.4 Cardiac computed tomography • 3.1 Coronary artery disease • 3.4 Coronary angiography • 7.5 Cardiac surgery

Learning points

- Intravascular imaging could identify the underlying mechanism and guide following intervention for patients with restenosis after direct reimplantation of anomalous origin of the left coronary artery from the pulmonary artery.
- It should be emphasized that regular follow-up is crucial for early recognition of this complication.
- Intensive secondary prevention, such as aggressive lipid-lowering therapy, should be considered to mitigate the risk of procedure failure.

Introduction

An anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rare congenital cardiac lesion with an estimated incidence of 0.2–1.2% of the general population.[1] In the presence of ALCAPA, the dependent myocardium is perfused by an anomalous coronary artery, which generally leads to the development of myocardial ischaemia, left ventricular dysfunction, and

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dilatation and mitral regurgitation (MR).[2,3] Long-term survival of adults with ALCAPA is possible because of the large number of compensatory collateral branches between the LCA and the right coronary artery (RCA); however, adults with ALCAPA may develop varying degrees of myocardial ischaemia.

Surgery is the first-line of treatment for ALCAPA, and the strategies mainly involve direct reimplantation of the anomalous coronary artery to the aorta, coronary button transfer in combination with the Lecompte manoeuvre, and extrapulmonary and intrapulmonary tunnelling techniques.[3] However, the long-term prognosis of this disease remains uncertain. Only a few cases of stenosis after the direct repair of ALCAPA have been reported so far, but underlying mechanism and management strategy remain unclear.

We have reported herein a case of ostial stenosis of a reimplanted left main coronary artery (LMCA) in a 15-year-old girl with ALCAPA. Subsequently, percutaneous coronary intervention (PCI) was performed and optimized under intravascular ultrasound (IVUS) guidance, and the patient's cardiac function improved gradually with significant relief of the ischaemic symptoms during the 5-year follow-up observations.

Timeline

Timeline	Clinical events
12 years old (1 year prior to surgery)	The patient presented with exertional angina pectoris. Echocardiography indicated reduced systolic function with left ventricular ejection fraction (EF) of 47%.
13 years old (day of surgery)	The patient was diagnosed with anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) by coronary computed tomography angiography (CCTA). Direct reimplantation of the anomalous coronary artery to the aorta was performed.
14 years old (3 months after surgery)	The patient complained recurrent retrosternal pain. Echocardiography showed normal systolic left ventricular functional with EF of 55.7%.
15 years old (day of PCI)	Coronary angiography and intravascular ultrasound (IVUS) revealed a restenosis of the left main coronary artery. A drug-eluting stent was implanted with IVUS guidance and a dual anti-platelet therapy was started.
15 years old (5 months after PCI)	The cardiac function improved. The coronary angiography showed that the LCA stent was patent without restenosis at the 5-month follow-up.
17 years old (2.5 years after PCI)	The cardiac function improved further. The CCTA demonstrated that the LCA stent was still patent without restenosis at the 2.5-year follow-up.
20 years old (5 years after PCI)	The patient was asymptomatic with preserved systolic function, in absence of residual coronary stenosis at the 5-year follow-up.

Case presentation

A 15-year-old girl of the Han nationality presented with exertional angina pectoris. She reported no relevant medical history, no smoking or drinking history and no family history of heart diseases. She was also not exposed to drugs, toxins, or allergens. Her echocardiography revealed a left ventricular end-diastolic diameter (LVEDD) of 5.7 cm, and the thickness of the left ventricular posterior wall and interventricular septum at end-diastole was 8.8 mm, while the left ventricular ejection fraction (EF) was 47%. The patient's cardiac magnetic resonance imaging (CMRI) revealed myocardial oedema and delayed enhancement of the local subendocardium and epicardium in the inferior basal wall, posterior wall, anterior wall and middle posterior wall of the left ventricle. Coronary computed tomography angiography (CCTA) showed the LCA originating from the pulmonary artery trunk, the presence of an LMCA–pulmonary artery trunk shunt, the establishment of multiple collateral circulations from the RCA to the left anterior descending artery branch and circumflex branch, and compensatory dilatation of the RCA (*Figure 1*). Direct reimplantation of the anomalous coronary artery to the aorta was performed with the support of extracorporeal circulation. The patient accordingly received medical therapy, including aspirin, a beta-blocker and isosorbide mononitrate sustained-release tablets after surgery. She was then prescribed with dihydrochlorothiazide and spironolactone for 1 month.

After 3 months of ALCAPA repair, the patient complained recurrent precordial pain. She showed no signs of heart failure, and her myocardial enzyme and brain natriuretic peptide (BNP) concentrations were normal. Her electrocardiography revealed left ventricular high voltage (R $wave_{(V5)}$ + S $wave_{(V1)}$ >3.5 mv), 0.1 mv ST-segment depression in the V3–V5 leads and T-wave inversion in the I, avL, and V2–V3 leads. Echocardiography demonstrated moderate to severe MR, LVEDD of 5.5 cm and the EF of 55.7%. CCTA demonstrated severe stenosis at the ostial LMCA (*Figure 2*). Coronary angiography revealed 90% stenosis at the ostium of the LMCA as well as ectasia of the proximal circumflex artery (*Figure 3*). Pre-PCI IVUS showed an attenuated plaque with a large plaque burden at the ostial LMCA, with the minimum lumen area of 4.2 mm^2 (the lower limit of normal is 4.5 mm^2). Through the IVUS guidance, a drug-eluting stent (BuMA biodegradable sirolimus-coated stent, 4.0 × 10 mm; SINOMED, Tianjin, China) was implanted in the ostial LMCA, and the final IVUS revealed that the minimum lumen area of 13.4 mm^2 (*Figure 4*). The

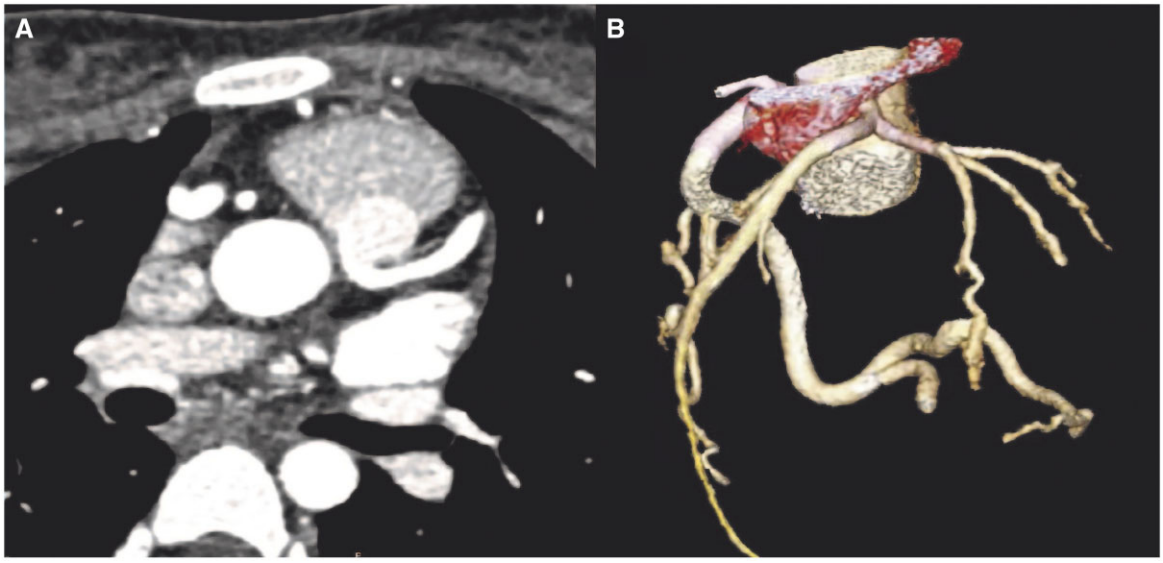

Figure 1 Two- and three-dimensional reconstructions of coronary computed tomography angiography (*A* and *B*) showing that the left coronary artery originated from the pulmonary artery trunk.

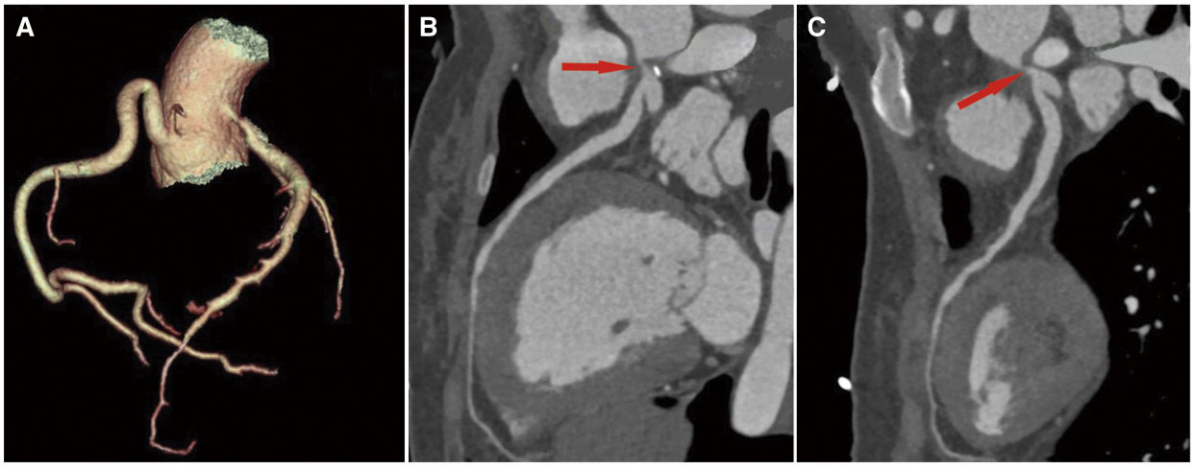

Figure 2 Two-dimensional (*B* and *C*) and three-dimensional reconstructions (*A*) of coronary computed tomography angiography showing severe stenosis at the left main coronary artery (red arrow).

patient accordingly received dual antiplatelet therapy (e.g. aspirin and clopidogrel), beta-blocker, isosorbide mononitrate sustained-release tablets, spironolactone, losartan potassium, trimetazidine dihydrochloride, and a proton pump inhibitor.

After 5 months of PCI, repeat echocardiography revealed moderate MR, LVEDD of 6 cm and a normal EF of 56.2%. Coronary angiography showed that the LMCA was patent without restenosis (*Figure 5*). At 2.5 years after PCI, the levels of creatine kinase-MB, cardiac troponin I, and BNP were all normal. Echocardiography showed that the LVEDD had decreased to 5.8 cm, while the EF was 58.8%. CCTA (*Figure 6*) revealed that the LMCA stent was still patent without restenosis. Presently, at 5-year follow-up, the patient is still free of cardiovascular events and is continuing with the prescribed medical therapy, including aspirin, simvastatin, perindopril, metoprolol succinate sustained-release tablets, and trimetazidine.

Discussion

Timely diagnosis and early treatment are crucial for patients with ALCAPA. CMRI and CCTA are important diagnostic examinations for ALCAPA. In fact, it is important to differentiate ALCAPA from

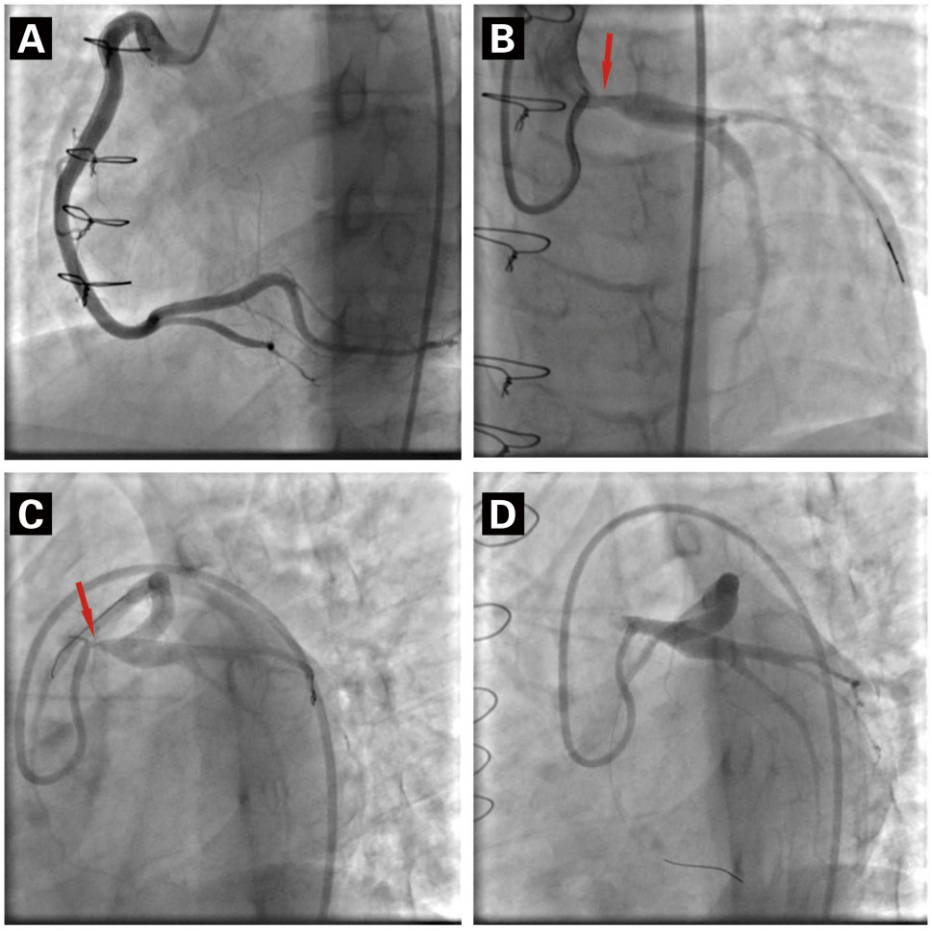

Figure 3 (*A*) Right coronary angiography showing no abnormalities. (*B* and *C*) Coronary angiography before percutaneous intervention (cranial view) indicating stenosis at the ostium of the left main coronary artery (red arrow). (*D*) Coronary angiography after percutaneous intervention.

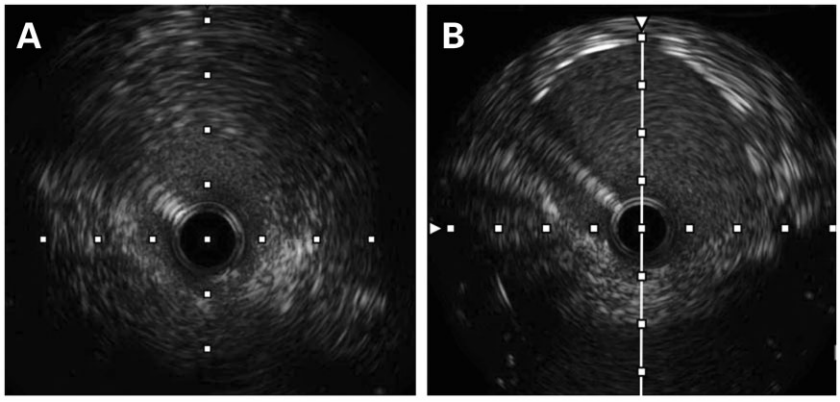

Figure 4 (*A*) Intravascular ultrasound before stent placement indicating severe stenosis at the ostium of the left main coronary artery, an attenuated plaque with a large plaque burden at the ostial left main coronary artery, and a minimum lumen area of 4.2 mm^2. (*B*) Intravascular ultrasound after stent placement suggesting that the minimum lumen area was 13.4 mm^2.

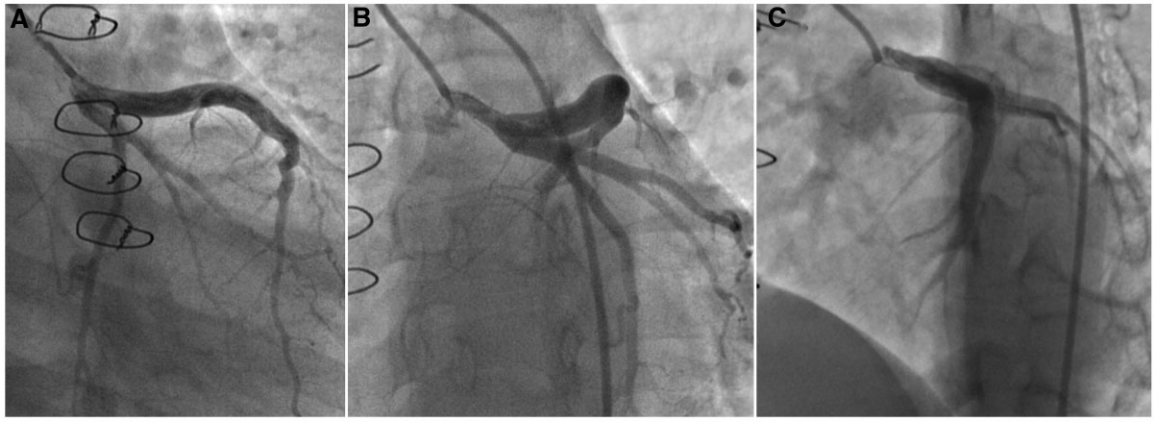

Figure 5 Coronary angiography after 5 months since the intervention (*A–C*) showing that the left main coronary artery was patent.

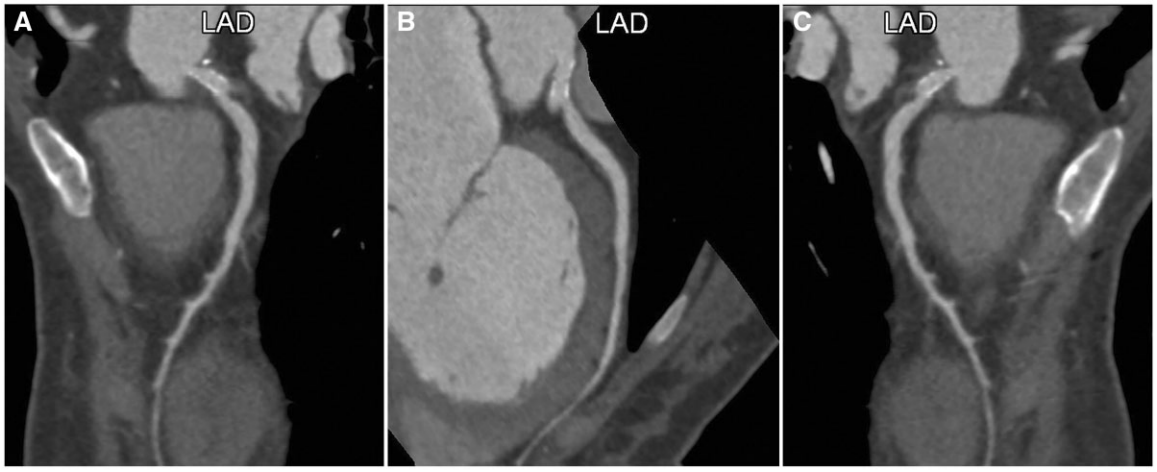

Figure 6 Coronary computed tomography angiography at 2.5 years after the intervention (*A–C*) showing that the left main coronary artery stent was still patent without restenosis. LAD, left anterior descending.

other diseases that cause coronary artery dilatation, including vasculitis (such as Kawasaki disease or arteritis), coronary-coronary sinus fistula, atresia, and atherosclerotic coronary artery dilatation.

All cases of ALCAPA require prompt surgical correction, considering that follow-up complications, such as arterial stenosis, persistent MR, and late-onset congestive heart failure may necessitate reinterventions.[4,5] Restenosis of a reimplanted LMCA is rare, with only a few cases reported so far.[6,7] The pathophysiological mechanism is ambiguous but may involve the long distance between anatomical structures, excessive extension or even distortion of the LCA, increased tissue stiffness, and metabolic disorders.[8] In fact, it has been hypothesized that the tractive force exerted on the LMCA after reimplantation and vascular rupture during an operation play the key role.[6,9] In the present case, pre-PCI IVUS demonstrated an attenuated plaque with a large plaque burden, suggesting that early atherosclerosis was accelerated after PCI and that any secondary preventive step may have been inadequate under the situation. In addition, it is possible that intimal injury and local hemodynamic disturbance related to surgery accelerated the process of ostial stenosis of the reimplanted LMCA. Importantly, regular follow-up needs to be emphasized as being crucial for early recognition of this complication. Moreover, more attention needs to be paid to intensive secondary prevention, such as through aggressive lipid-lowering therapy. Furthermore, we should make maximum efforts towards minimizing chances of intraoperative injury to the coronary artery.

The long-term clinical outcome of PCI remains unestablished. A past case report supports that PCI is a reasonable choice for patients at a high risk of reoperation-related complications.[6] Several past studies have confirmed that IVUS-guided coronary angiography can reduce the incidence of adverse cardiac events.[10,11] However, only a few reports have so far described the application of IVUS-guided PCI to treat stenosis of a reimplanted LMCA in patients with ALCAPA

without any long-term follow-up.[7] In the present case, optimal angiography and IVUS results were obtained, and the patient developed no significant symptoms and had relatively normal cardiac function. This outcome confirms the feasibility of IVUS-guided PCI in such patients.

Conclusion

We described the case of a 15-year-old girl with restenosis after direct reimplantation of ALCAPA. Our observations and experience suggest that atherosclerosis might be the mechanism for stenosis in such cases. IVUS-guided PCI can indeed be safely performed to provide a favourable midterm clinical outcome. Further studies are however warranted to better comprehend the mechanism of restenosis and explore the optimal management approach for this rare complication.

Lead author biography

Bo Zheng is currently a chief physician and a deputy director of the Institute of Cardiovascular Disease in Peking University First Hospital. He mainly engaged in percutaneous coronary interventional, intravascular imaging, and coronary physiology. He is currently fellow of European Society of Cardiology and fellow of American College of Cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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