ESC European Society of Cardiology

European Heart Journal - Case Reports (2023) **7**, 1–5
https://doi.org/10.1093/ehjcr/ytad230

CASE REPORT
Adult congenital heart disease

Ostial stenosis of reimplanted left main coronary artery and supravalvular pulmonary stenosis: a case report of two complications of surgery for anomalous left coronary artery from the pulmonary artery

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Received 4 October 2022; first decision 21 December 2022; accepted 28 April 2023; online publish-ahead-of-print 2 May 2023

Background Anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) is a rare congenital disease. Surgical re-implantation of the left main coronary artery (LMCA) to the aorta is a definitive treatment with a good prognosis.

Case summary A 9-year-old boy was admitted with a complaint of exertional chest pain and dyspnoea. At 13 months of age, he was diagnosed to have ALCAPA as a workup of severe left ventricular systolic dysfunction and underwent coronary re-implantation of ALCAPA. Coronary angiogram displayed the high takeoff of re-implanted LMCA with significant ostial stenosis, and echocardiogram showed significant supravalvular pulmonary stenosis (SVPS) with a peak gradient of 74 mmHg. After a multidisciplinary team discussion, he underwent percutaneous coronary intervention with stenting to ostial LMCA. On follow-up, he was asymptomatic and a cardiac computed tomography scan showed a patent stent in LMCA with an under-expanded area in the mid-segment. The proximal part of the LMCA stent was located very close to the stenotic segment of the main pulmonary artery making it a high risk for balloon angioplasty. The surgical intervention of SVPS is delayed to allow the somatic growth of the patient.

Discussion Percutaneous coronary intervention in re-implanted LMCA is a feasible option. If stenosis of re-implanted LMCA is accompanied by SVPS, the latter can be best treated surgically and staged to decrease the operative risk. Our case also demonstrates the importance of long-term follow-up of post-operative complications of patients with ALCAPA.

Keywords Case report • Anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) • Left main coronary artery (LMCA) • Supravalvular pulmonary stenosis (SVPS) • Percutaneous coronary intervention (PCI)

ESC Curriculum 2.2 Echocardiography • 2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease

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Handling Editor: Filippo Puricelli
Peer-reviewers: Jan Henzel; A Shaheer Ahmed; David Blusztein
Compliance Editor: Plyvios Demetriades
Supplementary Material Editor: Niklas Schenker

Learning points

- Stenosis of the re-implanted left main coronary artery (LMCA) and supravalvular pulmonary stenosis are complications of corrective surgery of anomalous origin of the coronary artery from the pulmonary artery.
- Stenting of stenosed re-implanted LMCA is a feasible option.
- Close follow-up with multimodality imaging by the multidisciplinary team is needed in such complicated cases.

Introduction

Anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) is a rare congenital disease. Surgical re-implantation of left main coronary artery (LMCA) to the aorta is a definitive treatment in such cases with a good prognosis.[1] Obstruction and stenosis of re-implanted LMCA are uncommon, and there is no consensus on its treatment strategies as experience is only limited to a few case reports. After corrective surgery, about one-third (24%) of patients experience supravalvular pulmonary stenosis (SVPS) at some stage.[2] Here, we present a case of a 9-year-old boy who underwent re-implantation of ALCAPA at the age of 13 months and now presented with exertional chest pain and dyspnoea.

Timeline

At 1 year of age (March 2012)	• Diagnosis of severe left ventricular systolic dysfunction • Diagnosis of ALCAPA • Corrective surgery for ALCAPA (re-implantation of LMCA to the aorta)
At 2 years of age (March 2013)	• Normalization of left ventricular systolic function
At 4 years of age (2015)	• Development of mild SVPS • Gradient across SVPS = 34 mm/Hg
At 9 years of age (2020)	• Exertional chest pain • Diagnosis of ostial stenosis of re-implanted LMCA • Severe SVPS (peak gradient = 75 mm/Hg) • Stenting of ostial LMCA
At 10 years of age (2021)	• A cardiac computed tomography (CT) scan showed the patent LMCA stent • The close proximity of SVPS and LMCA stent • The heart team consensus for staged surgery of SVPS
At 11 years of age (2022)	• Asymptomatic • Follow-up

Case presentation

A 9-year-old boy [weight = 35 kg, body mass index (BMI) = 17.9 kg/m^2, and body surface area (BSA) = 1.2 m^2] was electively admitted for workup after he complained of exertional chest pain and dyspnoea during his outpatient visit. There were no other associated symptoms. At 13 months of age, he was diagnosed to have ALCAPA as a workup of severe left ventricular systolic dysfunction and underwent corrective surgery with coronary re-implantation of ALCAPA. Initially, he was started on heart failure medications which were stopped after complete recovery of left ventricular function.

On clinical examination, he was vitally stable with a pulse rate of 84 beats/min and blood pressure of 112/62 mm/Hg. He was afebrile, and oxygen saturation at room air was 98%. Cardiac examination revealed the normal character of the first and second heart sound with an ejection systolic murmur at the left upper sternal border. His electrocardiography showed normal sinus rhythm with incomplete right bundle branch block with ST-segment depression and T-wave inversion in inferior leads (*Figure 1*). Transthoracic echocardiography revealed a mildly dilated right ventricle (RV) [proximal right ventricular outflow tract (RVOT) = 24 mm and RV base = 30 mm] with normal systolic function with mild RV hypertrophy. There was significant SVPS with a peak gradient of 74 mmHg. The left ventricle was normal in size (left ventricular internal diameter end diastole [LVIDd] = 36 mm, left ventricular internal diameter end systole [LVIDs] = 20 mm, and ejection fraction 60%) and functions without any regional wall motion abnormalities (*Figure 2* and Supplementary material online, *Video S1*). The diagnostic left and right heart catheterization showed significant SVPS (peak gradient 75 mm/Hg) with normal size and distribution of pulmonary artery branches (see Supplementary material online, *Video S2*). A non-selective angiogram showed the high takeoff of re-implanted LMCA with significant ostial stenosis with dilatation of the shaft (post-stenotic dilatation).

The patient was offered surgical intervention but the family was reluctant and requested alternate percutaneous therapy. After a multidisciplinary team discussion between adult and paediatric interventional cardiologists and cardiothoracic surgeons, the decision was made to proceed with further assessment of the LMCA followed by the percutaneous coronary intervention (PCI) and staged balloon dilatation of supravalvular stenosis.

During the percutaneous intervention under general anaesthesia, a 6 Fr femoral sheath was inserted with ultrasound guidance and then semi-selective imaging with contrast confirmed the presence of significant disease in the ostial LMCA (see Supplementary material online, *Video S3*). The intravascular ultrasound (IVUS) showed severe luminal area reduction at the ostium due to a fibrous band without any evidence of atherosclerosis (*Figure 3*). After pre-dilatation using semi-compliant and non-compliant balloons, a 4.5 × 12 mm Onyx drug-eluting stent (DES) was implanted and post-dilated with a non-compliant balloon (5.0 × 8 mm) at 20 atm (*Figure 4* and Supplementary material online, *Video S4*). The IVUS images showed good expansion and apposition with a minimum luminal area of 11 mm^2 at the ostium. The available stent with the best radial strength was chosen to avoid recoil. The patient was started on dual antiplatelet therapy (aspirin 81 mg daily and clopidogrel 75 mg daily).

On follow-up, the patient was asymptomatic. A repeated echocardiogram showed normal left ventricular systolic function (LVIDd = 38 mm, LVIDs = 21 mm, and ejection fraction 60%) and mildly dilated RV (proximal RVOT = 23 mm and RV base = 31 mm) with normal function. The supravalvular pulmonary gradient was stable (around 75 mm/Hg). About 10 months after the PCI, a cardiac CT was done to assess suitability for pulmonary valvuloplasty. It showed a patent stent in LMCA with an under-expanded area in the mid-segment (*Figure 5*). The proximal part of the LMCA stent was located very close to the stenotic segment of the main pulmonary artery (*Figure 5*). The case was discussed in the heart team meeting again and due to the

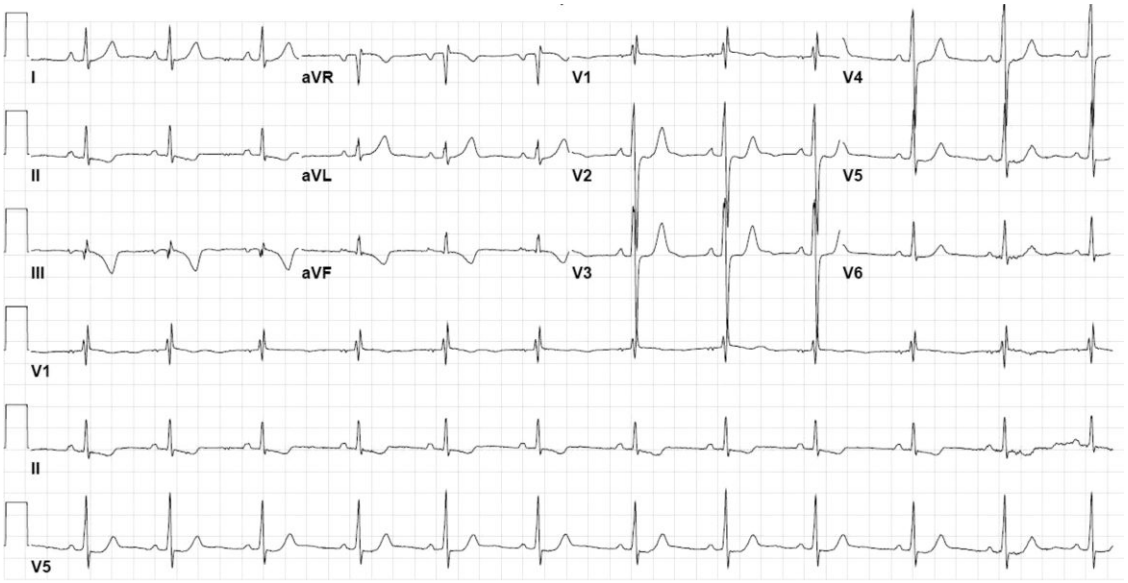

Figure 1 Electrocardiography of the patient showing normal sinus rhythm with incomplete right bundle branch block with ST-segment depression and T-wave inversion in inferior leads (leads II, III, and aVF).

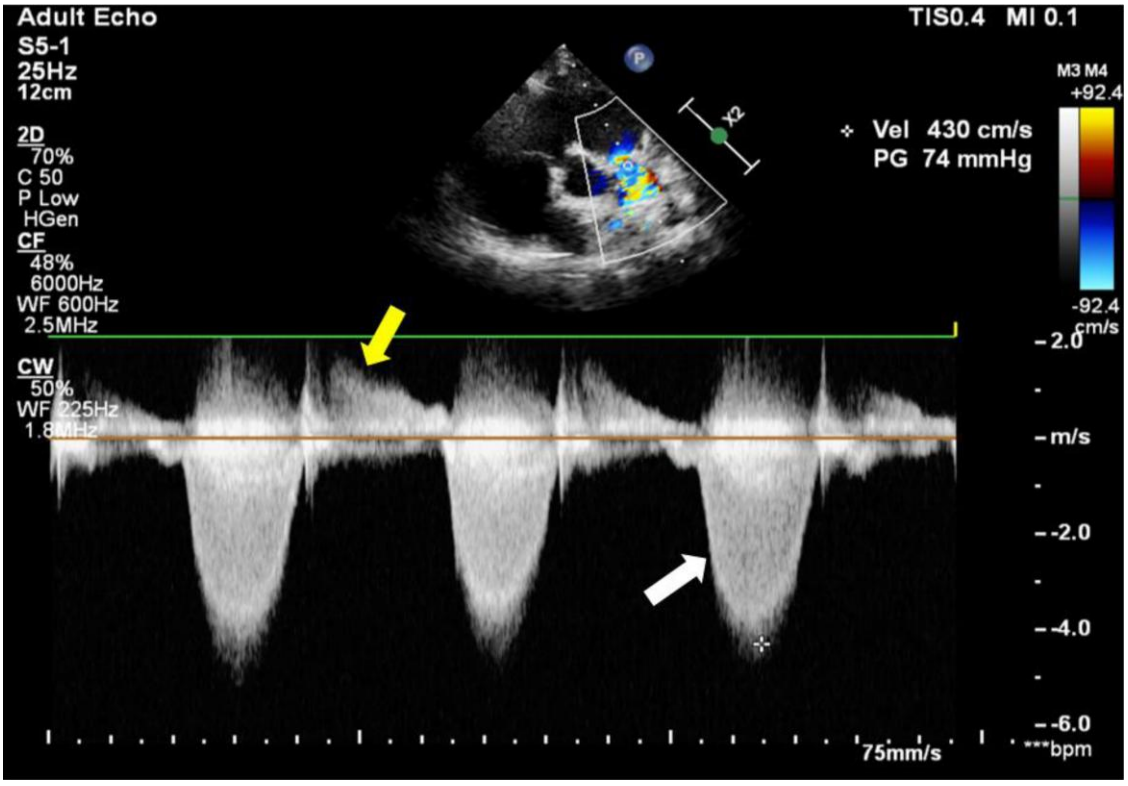

Figure 2 Continuous wave Doppler across the pulmonary valve and pulmonary artery showed a peak systolic gradient of 75 mm/Hg (white arrow) with a mild diastolic jet of pulmonary regurgitation (yellow arrow). CW, continuous wave.

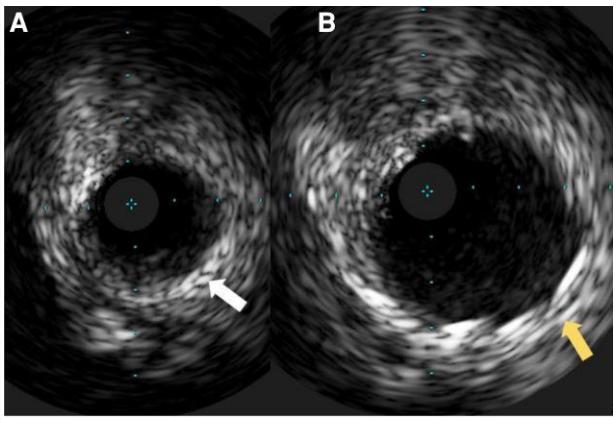

Figure 3 Intravascular ultrasound images at the ostium of the left main coronary artery showing a fibrous band (white arrow) causing severe luminal stenosis before the procedure (*A*) followed by stent implantation (yellow arrow) with adequate expansion (*B*).

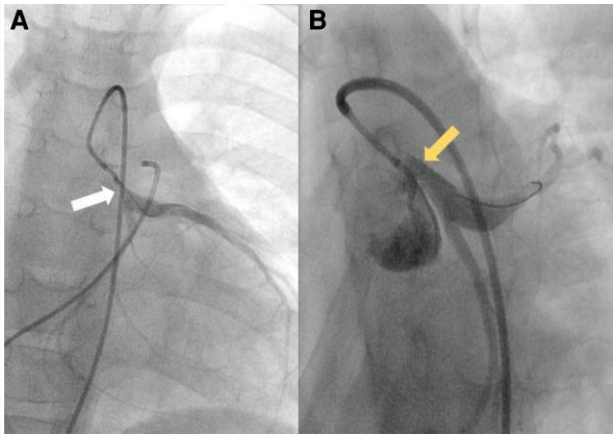

Figure 4 Cardiac catheterization still images pre- and post-stenting. (*A*) is a diagnostic injection to the left main coronary artery showing severe ostial stenosis (white arrow), and (*B*) shows the final image after stenting (yellow arrow).

proximity of the LMCA stent to the suprapulmonary stenotic area, the percutaneous angioplasty of SVPS was considered high risk due to the high likelihood of compression of the LMCA stent during the procedure. Currently, the patient is completely asymptomatic and under regular follow-up, and surgical intervention of SVPS is delayed to allow the somatic growth of the patient.

Discussion

Anomalous origin of the coronary artery from the pulmonary artery is a rare but life-threatening condition. Surgical re-implantation is considered an ultimate treatment with good long-term outcomes as other techniques such as coronary artery ligation, Takeuchi procedure, and

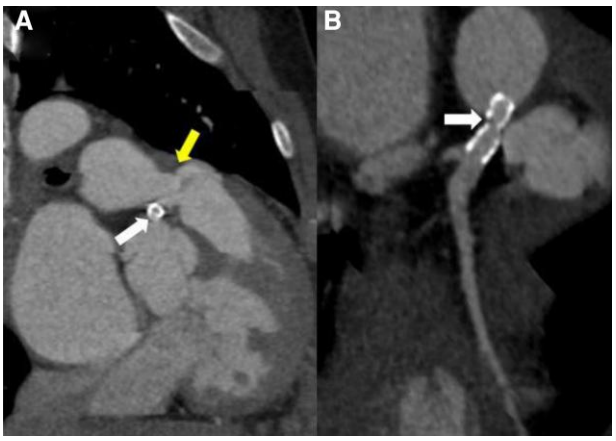

Figure 5 Coronal section of the cardiac computed tomography scan (*A*) showing the proximity of the left main coronary artery stent (white arrow) and supravalvular pulmonary stenosis (yellow arrow). Multiplanar reconstruction image (*B*) showing the patent stent in the left main coronary artery with some under expansion at the ostial site (note that the proximal edge of the stent is hanging into the aorta).

anastomosis to the subclavian artery have not proven to be superior in long-term outcomes.[1,2,3] Partial recovery of left ventricular function, persistence or worsening of mitral regurgitation, stenosis of re-implanted LMCA, and SVPS are some of the known complications of corrective surgery of ALCAPA.[1,4,5,6] The SVPS is a known complication (24%) after intra-pulmonary baffle (known as the Takeuchi procedure) and it is not commonly reported in direct re-implantation technique.[2] The exact incidence of stenosis of the re-implanted left coronary artery after corrective surgery of ALCAPA is not known due to limited data.[7,8] Our case presented with severe ostial stenosis of re-implanted LMCA and SVPS. To our knowledge, this is the first reported case of two late complications in one patient. The most common site of stenosis of re-implanted LMCA is the ostial segment. The exact mechanism is not known but likely the re-implanted segment of the coronary artery (ostial segment) heals with fibrotic tissue which is inelastic and prevents future growth causing stenosis.[9] Redo surgery is a reasonable option but it carries the risks associated with the second surgery, and the long-term outcomes of bypass grafting at a such young age are also unknown.

Percutaneous therapy with the use of stenting is an alternate option with some clinical and technical challenges. The smaller and growing size of coronaries in young patients, significantly less experience with coronary interventions in this age group, the possibility of artery recoil after stenting, and compliance with antiplatelet therapies are some of the challenges for percutaneous therapy.[8] Although good long-term outcomes of coronary stenting are reported in children, a retrospective analysis of 33 patients undergoing stent implantation due to various indications showed that almost three-fourths (73%) of them ended up in at least one re-dilatation, which highlights the need for close follow-up.[10,11] The DES with higher radial strength should be preferred over bare-metal stents (BMS) as they reduce the incidence of in-stent restenosis (ISR) by preventing neointimal hyperplasia. The treatment of SVPS is indicated in symptomatic or severe stenosis (defined by gradients more than 50 mm/Hg). The use of percutaneous modalities is limited by higher rates of restenosis due to vessel recoil after ballooning and the risk of thrombosis and migration after stenting.[12] Corrective surgery is the treatment of choice due to better long-term outcomes.[13]

In summary, PCI in re-implanted LMCA is a feasible option. The close collaboration of paediatric and adult cardiologists and cardiac surgeons is crucial in such complicated cases. If stenosis of re-implanted LMCA is accompanied by SVPS, the latter can be best treated surgically and staged to decrease the operative risk. Our case also demonstrates the importance of long-term follow-up of post-operative complications of patients with ALCAPA.

Lead author biography

Dr Abdullah Alkhodair is a consultant in interventional cardiology, structural heart diseases, and pulmonary hypertension and the adult cardiology program director at King Fahad Medical City, Riyadh, Saudi Arabia. He graduated with honours from King Saud University, Riyadh. He completed his internal medicine and cardiology training in Canada and holds the American Board of Internal Medicine, and cardiovascular diseases, and is certified by the Royal College of Physicians and Surgeons of Canada Certificate in internal medicine and cardiology. He did a fellowship in pulmonary hypertension followed by interventional and structural heart disease fellowship at the University of British Columbia.

Supplementary material

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

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 the submission and publication of this case report, including images and associated text, has been obtained from the patient and family in line with the COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

Data availability

The data underlying this article are available in the article and in its online supplementary material.

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