ESC European Society of Cardiology
European Heart Journal - Case Reports
doi:10.1093/ehjcr/ytab074

CASE REPORT
Congenital heart disease

Severe baffle leak after Takeuchi repair successfully treated with coronary bypass and percutaneous baffle closure: a case report

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Received 5 July 2020; first decision 2 September 2020; accepted 10 February 2021

Background Anomalous left coronary artery from the pulmonary artery is a rare congenital abnormality that requires surgical correction.

Case summary We describe the case of a 33-year-old female with a history of anomalous left coronary artery of the pulmonary artery who presents with exertional angina. She underwent a Takeuchi repair that was complicated by a baffle leak. She was successfully treated with left internal mammary artery-left anterior descending (LAD) bypass grafting and percutaneous baffle leak closure.

Discussion The Takeuchi procedure involves the creation of an aortopulmonary window and an intrapulmonary tunnel that 'baffles' the aorta to the ostium of the anomalous left coronary artery. The most common late complication of the Takeuchi procedure is the presence of a baffle leak. Percutaneous baffle leak occlusion via vascular plug and coronary bypass of the LAD can successfully treat a baffle leak with excellent short-term follow-up.

Keywords Case report • Takeuchi procedure • Baffle leak • Coronary bypass • Vascular plug

Learning points

- Anomalous left coronary artery from the pulmonary artery is a rare congenital abnormality that results in high mortality and requires surgical correction.
- The Takeuchi procedure involves the creation of an aortopulmonary window and an intrapulmonary tunnel that 'baffles' the aorta to the ostium of the anomalous left coronary artery.
- The most common late complication of the Takeuchi procedure is the presence of a baffle leak and supravalvular pulmonary stenosis.
- These complications may be repaired surgically although this often involves risk from repeat thoracotomy or using a percutaneous approach.

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Handling Editor: Christoph Sinning
Peer-reviewers: Ina Michel-Behnke and Inga Voges
Compliance Editor: Hibba Kurdi
Supplementary Material Editor: Mariame Chakir

Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality that requires surgical correction. There are two types of ALCAPA syndrome, the infant type and the adult type.[1] Both of these subtypes of ALCAPA have presenting features and outcomes. The infant type manifests as myocardial infarction and congestive heart failure, and approximately 90% die within the 1st year of life. The adult type is rare but can cause angina, congestive heart failure, or sudden cardiac death. In this case, we describe the case of an adult who presents with symptomatic ALCAPA syndrome.

Timeline

Day 0	**Presentation to previous institution with complaints of exertional angina and dyspnoea after delivery of her first child. She was diagnosed with post-partum cardiomyopathy and ALCAPA syndrome. She subsequently underwent Takeuchi repair.**
Month 7	**She presented to our institution and computed tomography coronary angiogram revealed a 6.5 x 2.4mm leak present from the distal inferior Takeuchi baffle to the main pulmonary artery as well as a smaller leak near the baffle origin. After multi-disciplinary discussions, she underwent successful LIMA-LAD bypass with percutaneous baffle closure with Amplatzer Vascular Plug II.**
Month 31	**Re-presented with complaints of angina. Cardiac catheterization revealed a patent RCA, persistent occlusion of the Takeuchi baffle by the vascular plug without any leaks, a patent LIMA graft to LAD, however, there was a chronic total occlusion of the proximal left circumflex artery with collaterals. Her angina was medically managed successfully.**

Case description

A 33-year-old white Caucasian woman with a past medical history of peripartum cardiomyopathy, ALCAPA corrected with a Takeuchi procedure, who presented to her cardiologist's office with progressive dyspnoea, fatigue, and exertional angina. The Takeuchi repair is a surgical procedure that involves the creation of an aortopulmonary (AP) window and an intrapulmonary tunnel that 'baffles' the aorta to the ostium of the anomalous left coronary artery. Our patient's clinical history was unremarkable for any cardiac issues in infancy, early childhood, and adolescence. She initially became symptomatic after the delivery of her first child. She was diagnosed with post-partum cardiomyopathy after her transthoracic echocardiogram revealed a moderately reduced left ventricular ejection fraction (LVEF). She was treated medically with metoprolol succinate 25 mg daily and Lisinopril 10 mg daily. The diagnosis of ALCAPA was made after her cardiac magnetic resonance imaging (MRI) demonstrated a dilated left ventricle, mild mitral valve regurgitation, regional hypokinesis of the mid-to-apical anterior wall, and mid-to-apical anterior septum. The patient underwent a Takeuchi repair 7 months prior at an outside institution due to exertional angina for 4 months. It was unclear why Takeuchi repair was chosen over other surgical techniques such as left coronary button transfer to the aorta or bypass grafting with proximal ligation of the anomalous artery. Her echocardiogram showed a moderately reduced LVEF of 40%. Her symptoms of angina and LVEF normalized after her surgery until 1 month prior to presentation.

She presented to our institution with complaints of exertional angina, shortness of breath, and leg oedema worsening over the last 2 months. Her vital signs were within normal limits. Her cardiac exam was positive for a biphasic continuous murmur, with a two out of six decrescendo systolic murmur at the left upper sternal border followed by a soft one out of four diastolic murmur. She did not have physical findings of volume overload. Her complete blood count, basic chemistry panel, liver function tests, cardiac troponins, and N-terminal prohormone of brain natriuretic peptide were within normal limits. A repeat cardiac MRI showed findings consistent with chronic left anterior descending (LAD) territory myocardial scar (Supplementary material online, *Figure S1*). Nuclear stress test revealed a large perfusion defect in the anterior wall with moderate superimposed ischaemia (Supplementary material online, *Figure S2*). Cardiac computed tomography (CT) coronary angiogram revealed a 6.5 mm × 2.4 mm leak present from the distal inferior Takeuchi baffle to the main pulmonary artery (*Figure 1A* and *B*) as well as a smaller leak near the baffle origin (not shown). Right heart catheterization confirmed a Qp:Qs of 1.5 with mild pulmonary hypertension. Coronary angiogram revealed right coronary artery (RCA) ectasia. Angiography of the baffle revealed marked left-to-right shunting from the baffle to the main pulmonary artery. The proximal LAD and left circumflex (LCx) arteries were patent but dilated because of the ALCAPA prior to Takeuchi repair. There was also minor fistulous connections present from the LAD to the left ventricle. After multidisciplinary consultation with cardiac surgery, adult congenital heart disease, and interventional cardiology services, the decision was made to proceed with a hybrid treatment approach of bypass of left internal mammary artery (LIMA) grafting to the LAD followed by

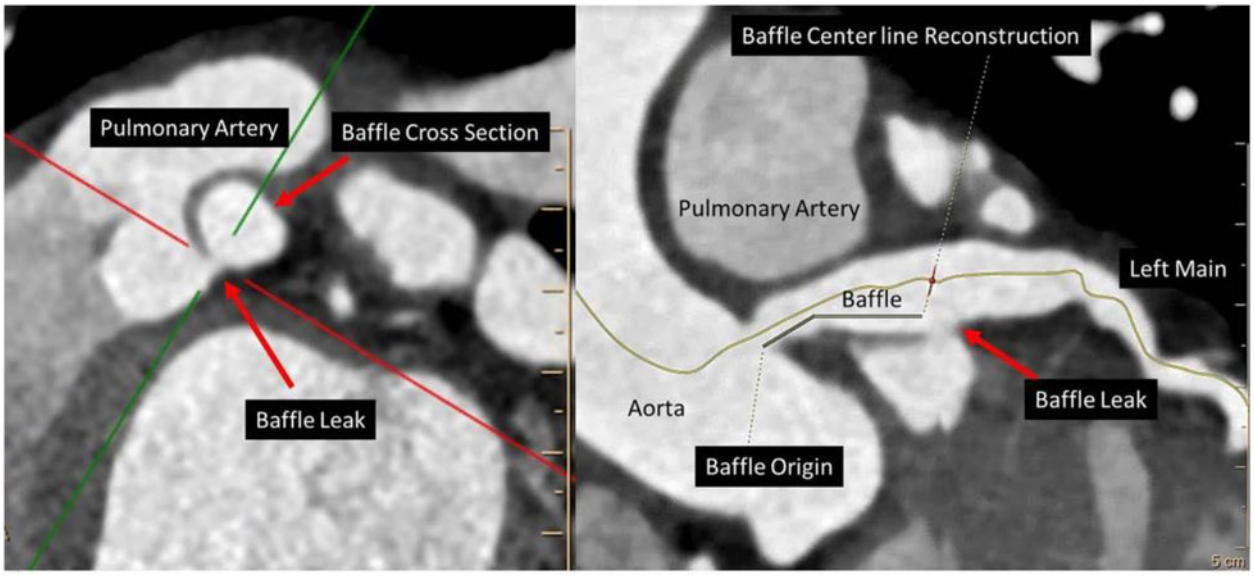


Figure 1 (*A*/*B*). A computed tomography coronary angiogram revealed a 6.5 mm × 2.4 mm leak present from the distal inferior Takeuchi baffle to the main pulmonary artery.

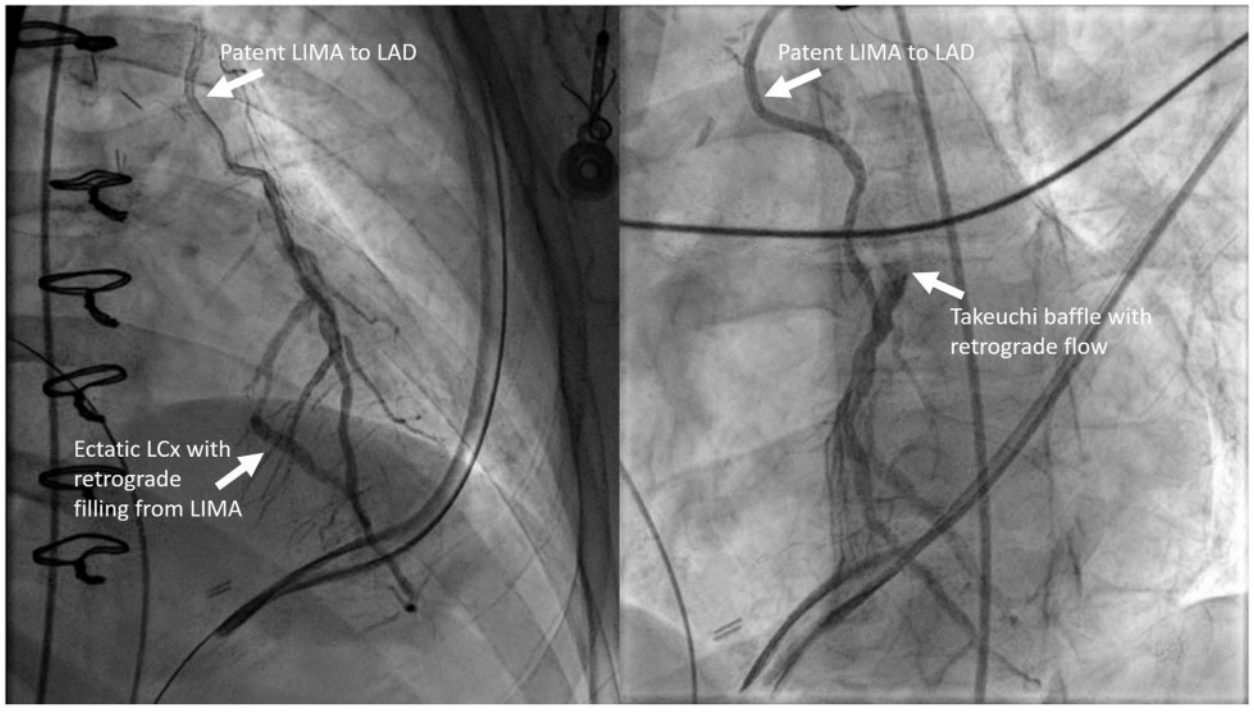


Figure 2 (*A*/*B*). Coronary angiography reveals left internal mammary artery-to-left anterior descending artery bypass graft patency and evidence of back filling into the baffle. LAD, left anterior descending artery; LIMA, left internal mammary artery.

percutaneous baffle embolization using the Amplatzer Vascular Plug II (Abbott, USA). Redo-cardiac surgery to repair the baffle was considered but felt to be high-risk. Percutaneous baffle leak closure was considered, but the presence of two leaks suggested weakness of the baffle suture line, which could be exacerbated by placement of closure devices.

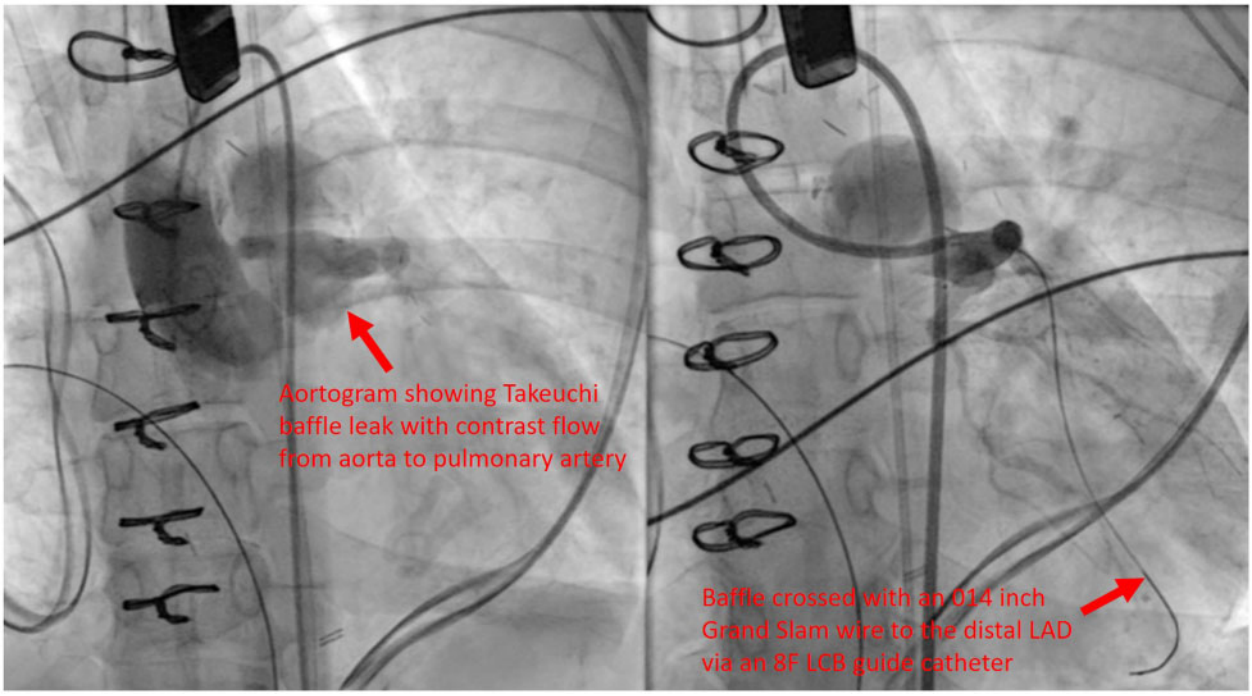


Figure 3 (*A*/*B*) Aortography shows the baffle with leakage of contrast into the pulmonary artery (left). The baffle was crossed with a Runthrough wire via an 8-Fr left coronary bypass guide catheter via the femoral artery (right).

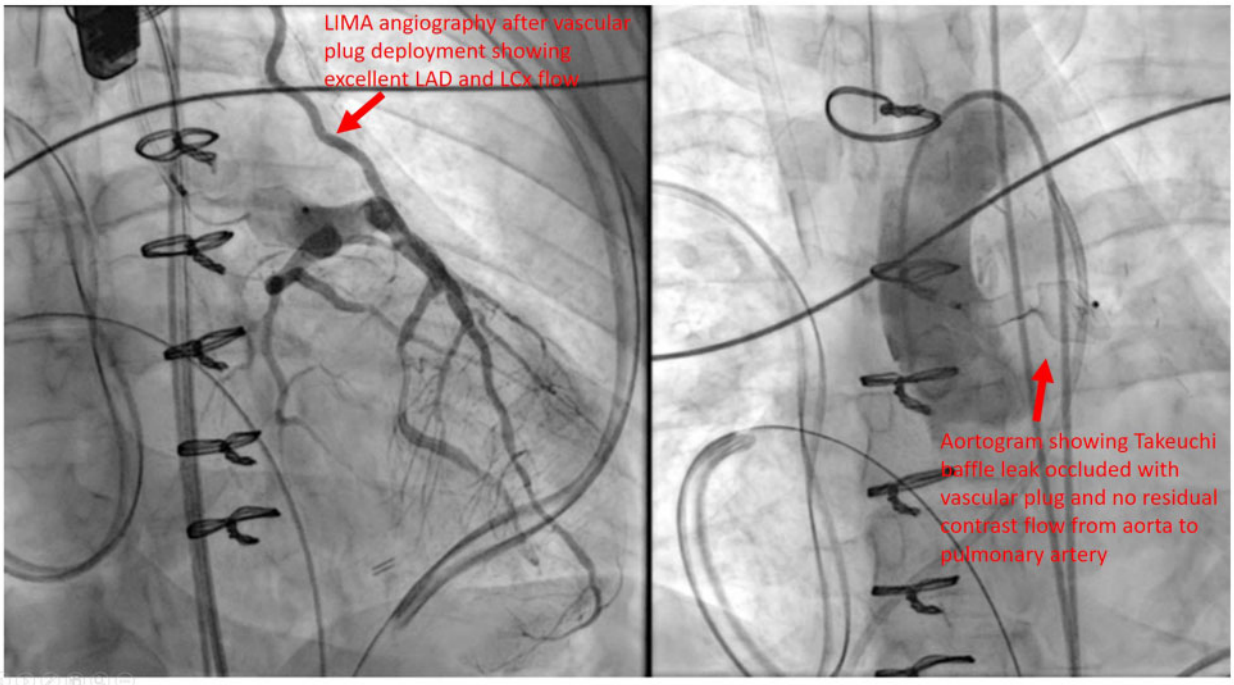


Figure 4 (*A*/*B*) A 14 mm Amplatzer Vascular Plug II was deployed from the left main coronary artery proximal to the bifurcation back to the aorta through the full extent of the baffle (left). Aortogram reveals no contrast flow into the pulmonary artery (right).

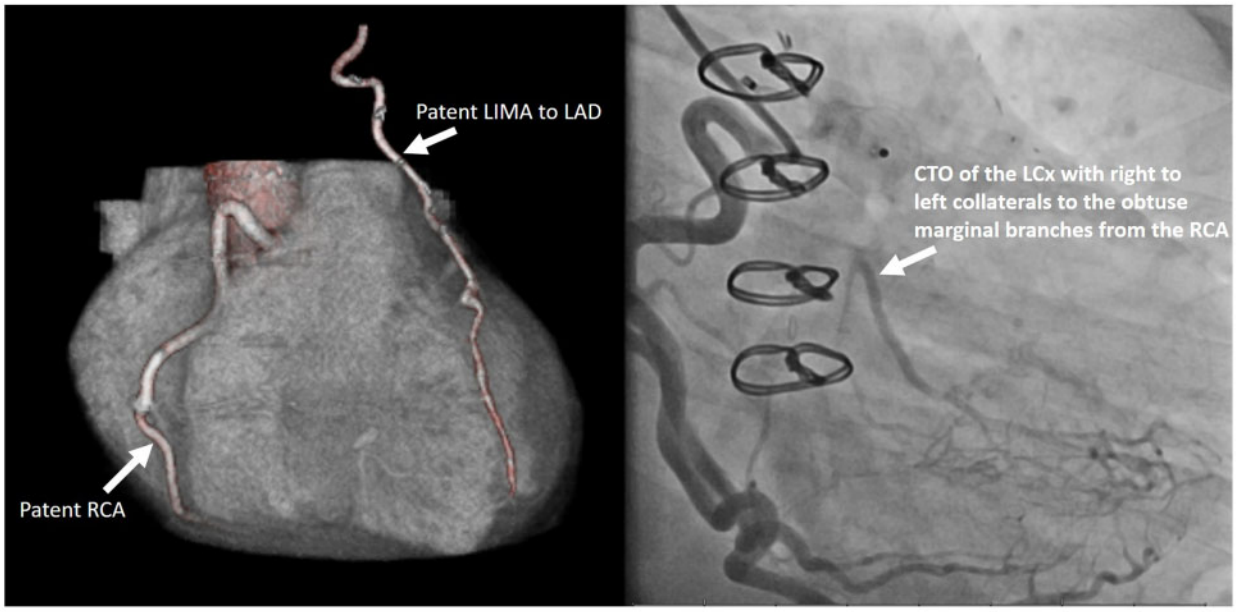
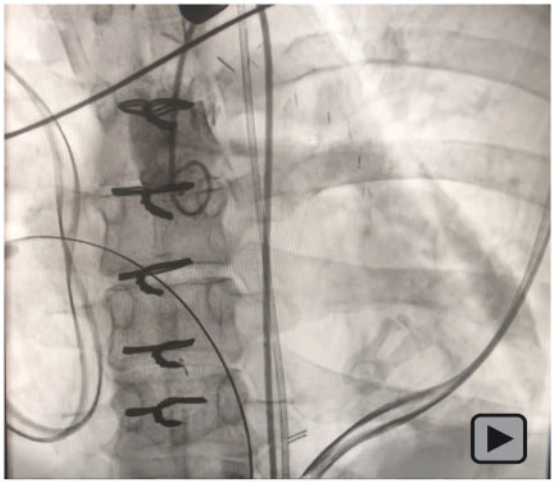


Figure 5 (*A*/*B*) A repeat coronary computed tomography angiogram revealed a patent right coronary artery and left anterior descending artery, but absent flow in the left circumflex artery (left). A repeat cardiac catheterization revealed persistent occlusion of the Takeuchi baffle by the vascular plugs without any leaks, patent left internal mammary artery to left anterior descending artery, patent right coronary artery and chronic total occlusion of the proximal left circumflex artery with right to left collaterals filling her obtuse marginal branches (right). CTO, chronic total occlusion; LAD, left anterior descending; LCx, left circumflex artery; LIMA, left internal mammary artery; RCA, right coronary artery.



Video 1 Angiogram of the LIMA to LAD is performed post-bypass. Then an aortogram was performed identifying the baffle leak. The baffle was crossed with an 014 inch Grand Slam wire to the distal LAD via an 8F LCB guide catheter. A vascular plug was deployed in the baffle and angiogram confirmed that the leak was sealed. After plug deployment, the LIMA was then engaged to confirm no backfilling of the baffle.

A LIMA-to-LAD bypass was performed followed by coronary angiography that showed graft patency with the back filling into the baffle (*Figure 2A and B*). An ascending aortogram was performed to identify the AP window origin of the baffle and confirmed the baffle leak with contrast leakage into the pulmonary artery (*Figure 3A*). Using a transfemoral artery approach, the baffle was engaged with an 8-Fr left coronary bypass coronary guide catheter, and the baffle crossed with a 0.014-inch Runthrough NS wire (Terumo, Japan) into the LAD. The Runthrough NS wire was exchanged over a Transit micro-catheter for a 0.014-inch Grand Slam wire (ASAHI, Japan) for better support (*Figure 3B*). A 14 mm Amplatzer Vascular Plug II was deployed from the left main coronary artery at the bifurcation back to the aorta through the full extent of the baffle. Repeat LIMA angiography (*Figure 4A*) and root aortography (*Figure 4B*) showed effective closure of both ends of the baffle without any leak into the pulmonary artery. The LIMA graft supplied the entire left coronary circulation. The patient recovered well post-operatively with resolution of her symptoms and normalization of her LVEF. Given the potential for thrombus formation presumably due to stagnant blood flow in ectatic coronary arteries with myocardial infarction which has been seen immediately after percutaneous coronary arteriovenous fistula embolization, oral anticoagulation with warfarin in addition to aspirin was started to prevent thrombosis in the ectatic left coronary artery which was now solely supplied by the LIMA graft. She took warfarin with a goal international normalized ratio (INR) of 2–3

for 4 months to allow some time for regression of vessel size at which point warfarin was discontinued.

Two-years following her surgery, she re-presented to her cardiologist's office with complaints of angina on moderate exertion with radiation to her left arm. Her angina was associated with shortness of breath, diaphoresis, and nausea and was relieved with rest. A repeat coronary CT angiogram revealed a patent RCA and LAD, but absent flow in the left circumflex artery (*Figure 5A*). There was no change in the configuration of the Vascular Plug II position or configuration compared to immediately following the procedure. Repeat cardiac catheterization revealed a patent RCA, persistent occlusion of the Takeuchi baffle by the vascular plug without any leaks, a patent LIMA graft to LAD, however, there was now a chronic total occlusion (CTO) of the proximal LCx artery with right to left collaterals filling her obtuse marginal branches (*Figure 5B*). In retrospect, it was thought that she likely had late thrombosis of her persistently ectatic left circumflex artery due to stagnant flow, now off of oral anticoagulation which was prescribed for only a short course following the procedure. She was started on medical therapy for her symptoms consisting of metoprolol succinate 50 mg once daily and isosorbide mononitrate 30 mg once daily with good effect. We opted to manage her angina medically and if she fails medical treatment, then to consider complex retrograde CTO percutaneous coronary intervention in the future. The patient remains asymptomatic from a cardiac standpoint after 8 months of medical therapy.

Discussion

Anomalous left coronary artery from the pulmonary artery is a rare congenital abnormality that results in high mortality if left untreated. The mechanisms underlying morbidity and mortality involve left ventricular dysfunction as well as progressive ischaemic valvular disease. Anomalous left coronary artery from the pulmonary artery requires surgical closure soon after it is diagnosed. In certain situations, the Takeuchi procedure is a viable alternative. The most common late complication of the Takeuchi procedure is the presence of a baffle leak. Other common complications are the presence of supravalvular pulmonary stenosis.[2] These complications may be repaired surgically although this often involves risk from repeat thoracotomy or using a percutaneous approach.[3] To our knowledge, this is the first case of Takeuchi baffle leak successfully treated with LIMA-LAD bypass grafting and percutaneous baffle leak closure.

Lead author biography

Syed Y. Naqvi is an Interventional Cardiology Fellow at the University of Rochester.

Supplementary material

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

Slide sets: A fully edited slide set detailing these cases 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.

Conflict of interest: None declared.

Funding: None declared.

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