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Reanastomosis and rehabilitation of interrupted left pulmonary artery derived from atretic ductus

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Brief Report

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Abstract

True absence of a branch pulmonary artery is rare. We identified a patient initially diagnosed with an absent left pulmonary artery at a previous hospital. Due to disagreement in the initial diagnosis, she had a diagnostic catheterisation, which revealed an isolated left pulmonary artery off the left innominate artery via a ductus. The ductus was recanalised with serial stenting and balloon dilatation followed by reanastomosis to the main pulmonary artery. In a patient who initially is diagnosed with an absent pulmonary artery, an alternative diagnosis, such as this case report, should be considered.

An absent branch pulmonary artery was first described by Frantzel in 1868.[1] It was later found in 1954 by McKim that true absence of a branch pulmonary artery is unusual and that there was typically a ductus arteriosus origin present during fetal life.[2,3] Surgical repair of a discontinuous branch pulmonary artery has been described as early as 1984 by Cobanoglu[4] and followed by others.[5,6] One surgical approach includes a Blalock–Taussig shunt to feed the hypoplastic branch artery. Alternatively, repair by stent recanalisation followed by surgical reanastomosis to the main pulmonary artery[7] can be considered; however, stenting of the ductus remains limited worldwide with mixed results.[8–11]

This report identified a patient with left lung hypoplasia who was initially diagnosed with absent left pulmonary artery at a previous hospital. In consideration of an alternative diagnosis, she underwent interventional catheterisation at our institution which revealed ductal origin of the left pulmonary artery via the left innominate artery. This report describes a staged approach using serial recanalisation of the ductus followed by reanastomosis to the main pulmonary artery.

Case presentation

An 8-month-old female with history of chronic colds, bronchospasm, and left lung hypoplasia presented to the cardiology clinic with a 3/6 holosystolic murmur at the left sternal border. She had been previously diagnosed with a moderate-restrictive ventricular septal defect, right-sided aortic arch, and absent left pulmonary artery on echocardiogram and cardiac CT angiogram at an outside hospital. She was lost to follow-up and subsequently referred to our institution several months later. Upon review of the cardiac CT, a subtle dimple on the left innominate artery was seen and suspicion arose for a ductal origin left pulmonary artery. The left pulmonary artery could not be visualised, nevertheless, a small left pulmonary vein could be identified on cardiac CT. She underwent heart catheterisation, which confirmed a left innominate artery dimple (Fig 1). Prior to the procedure, she was anticoagulated with aspirin and during the procedure received heparin 100 units/kg.

Initial catheterisation data showed elevated right pulmonary artery, right ventricle , and right atrial pressures. Pressures in the right pulmonary artery measured 37/18 mmHg with a mean of 27 mmHg. Main pulmonary artery demonstrated pressures of 43/18 mmHg with a mean of 30 mmHg. Pressures in the right ventricle measured 57/15 mmHg and the right atrial pressures measured A-wave of 17 mmHg, V-wave of 14 mmHg, and a mean of 12 mmHg.

Left pulmonary vein angiogram demonstrated reflux of contrast into left pulmonary artery to prove its patency as well as positioning prior to recanalisation of the innominate ductus. Recanalisation was performed using a 4-French Judkins right 2.5 coronary catheter, advanced from the right femoral artery into the left innominate artery. The catheter was able to engage the ductal dimple where an angiogram was performed and demonstrated scant flow from the dimple to left pulmonary artery. A Corsair microcatheter (Asahi Intecc USA Inc., Tustin, USA) and 0.014" chronic total occlusion guide wire (Asahi Intecc USA Inc., Tustin, USA) were advanced through the Judkins catheter to engage the ductal dimple. The wire was gradually advanced through the ductus into the distal left lower lobe pulmonary artery. An additional angiogram was performed to confirm contrast flowing from the ductal dimple into the left

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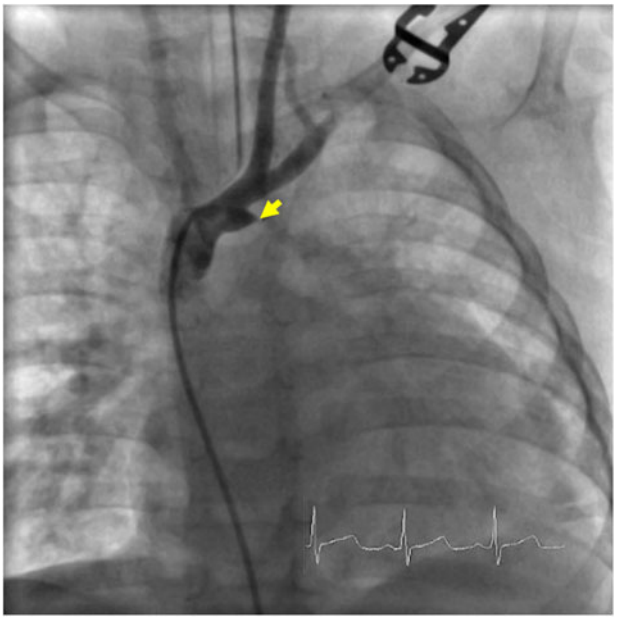

Figure 1. Selective innominate artery angiogram in antero-posterior projection demonstrated the ductus dimple (arrow).

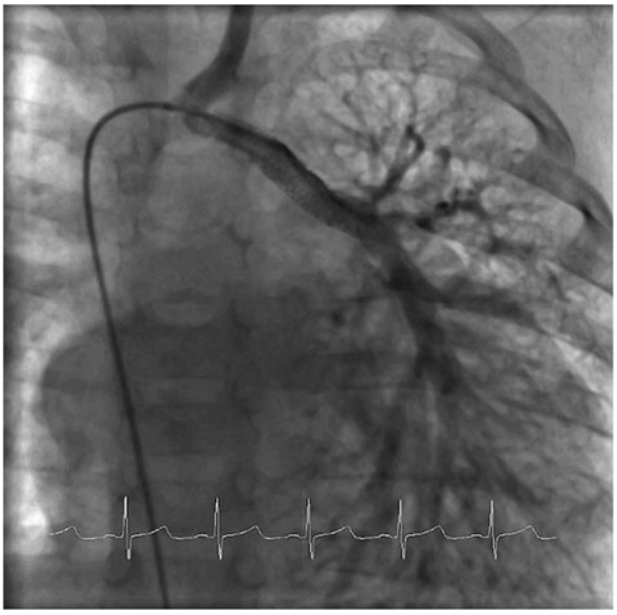

Figure 2. Selective innominate artery angiogram in antero-posterior projection after ductus stenting.

pulmonary artery on bi-plane. The microcatheter was advanced over the wire into the distal left lower lobe pulmonary artery, and the chronic total occlusion guide wire was removed. This was replaced with a 0.014" Ironman coronary guide wire (Abbott, Santa Clara, USA). The 4-French Judkins catheter and 5-French arterial sheath were then removed and replaced with a long 5-French Cook sheath which was advanced into the innominate artery. Using first a 2 mm × 20 mm coronary balloon catheter, followed by a 3 mm × 20 mm coronary balloon catheter, several inflations were performed along the length of the closed ductus. Subsequently, a 4 mm × 20 mm Rebel coronary stent (Boston Scientific, San Jose, USA) was advanced to straddle the distal stenosis of the ductus and inflated to 18 atm. A second Multi-Link Vision stent (Abbott, Santa Clara, USA) 4 mm × 18 mm was utilised to cover the remaining stenosed part of the ductus in an interlocking manner. Repeat angiograms showed the ductus to be fully open (Fig 2). Echocardiogram demonstrated a gradient of 17 mmHg with continuous flow through the stents. She remained receiving aspirin post-procedure.

At 16 months of age, balloon angioplasty of the ductal stents was performed secondary to ingrowth obstruction using a 5-mm coronary balloon catheter. The stents diameter increased to 5 mm with adequate continuous flow into the hypoplastic left pulmonary artery demonstrated by echocardiographic follow-up.

At 21 months of age, she underwent surgery for ventricular septal defect closure, ductus ligation, and left pulmonary arterioplasty. The stents were divided and extended to the hilum of the left pulmonary artery. The distal stent was removed. An incision was made to the main pulmonary artery in a door-shaped fashion and the left pulmonary artery was anastomosed to the main pulmonary artery using 6-0 Prolene sutures. A pericardial patch was placed to the anterior side to finish the anastomosis. Day 1 post-operative echocardiogram demonstrated patent flow through the left pulmonary artery with a peak gradient of 14 mmHg.

At 9-month post-operative follow-up, the patient was growing well without particular clinical concerns. Echocardiogram demonstrated the left pulmonary artery diameter at 4.5 mm with a peak gradient of 20 mmHg. Cardiac catheterisation follow-up at the age of 2.75 years demonstrated favourable improvements of the right pulmonary artery, the right ventricle, and the right atrial pressures. There was no pressure difference in the main pulmonary artery and the right pulmonary artery, both measured 27/9 (17) mmHg. The right ventricle pressure was 27/6 mmHg and the right atrial pressure was 8/6 mmHg. However, although the left pulmonary artery remained patent, it appeared considerably smaller than the right pulmonary artery with a pressure of 13/4 (9) mmHg and gradient of 14 mmHg. As a result, the patient received a second balloon angioplasty of the proximal left pulmonary artery resulting in improvement of the diameter and reduction of the pressure gradient (from 14 to 6 mmHg).

Discussion

An occluded ductus arteriosus arising from the first aortic branch to supply the left pulmonary artery causing left pulmonary artery interruption, although rare, has potential for transcatheter left pulmonary artery revascularisation. Diagnosis of ductal origin pulmonary artery is often delayed and challenged by the numerous anatomical variants and diverse clinical presentations.[12] Patients can present with recurrent respiratory infections and presumed pulmonary hypoplasia in the neonatal period or remain asymptomatic with insidious development of pulmonary hypertension and heart failure. The patient was initially diagnosed with an absent left pulmonary artery at an outside hospital, but after re-evaluation at our institution was correctly diagnosed following cardiac catheterisation. Cardiac catheterisation and pulmonary vein wedge angiography are diagnostic for ductal arising pulmonary artery by identification of the discontinuous hilar end of the pulmonary artery. Less invasive alternatives include cardiac CT and cardiac MRI, but are less sensitive if the ductus has closed.[13]

There are two broad strategies for resuscitation of ductal origin pulmonary artery: primary reanastomosis of ductal origin pulmonary artery to the main pulmonary artery and augmentation of distal pulmonary blood flow (via Blalock–Taussig shunt or ductal

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stenting) followed by reanastomosis. Primary reanastomosis has been recommended at earlier age of diagnosis. Authors theorise that younger patients are more likely to have a patent ductus with acceptable pulmonary blood flow, and that early reanastomosis will preserve and allow for distal pulmonary vascular growth.[12] When the diagnosis of ductal origin pulmonary artery is made later, ductal closure and resultant pulmonary artery hypoplasia may necessitate creation of a Blalock–Taussig shunt to improve pulmonary flow and vascular growth prior to reanastomosis. A less invasive alternative, if the ductus can be crossed by a wire, is the creation of a ductal stent through cardiac catheterisation. In this particular case, in which the descending aorta is on the right side and the duct comes from the left innominate artery, duct stenting is technically more straightforward and therefore we decided on this approach. However, the process of recanalisation can be more cumbersome. It is recommended prior to reanastomosis to stent to an internal diameter of at least 4–5 mm to mitigate risk of thrombosis and promote laminar flow.[14]

The diagnosis of ductal origin pulmonary artery should be considered in a patient with presumed pulmonary hypoplasia. This condition is often underrecognised because following closure of the ductus arteriosus, absence of flow to the affected pulmonary artery leads to misdiagnosis of an absent pulmonary artery.[15] Therefore, these patients are not offered correction. This report highlights the importance of considering ductal origin pulmonary artery in a patient who presents with presumed lung hypoplasia. This case demonstrates successful identification of and stent recanalisation of an isolated left pulmonary artery off the innominate artery leading ultimately to successful reanastomosis and patency.

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Conflicts of Interest. None

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