Catheter rehabilitation of occluded aberrant pulmonary artery

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Received 16 November 2012; received in revised form 16 January 2013; accepted 26 January 2013

Abstract

A 10-year old girl underwent a CT angiogram to investigate right lung hypoplasia. This showed a normal bronchial tree, lung parenchyma and pulmonary venous drainage, but an absent right pulmonary artery. Cardiac catheterization with pulmonary vein wedge injections identified a disconnected hypoplastic right pulmonary artery system supplied by an occluded right-sided ductus arteriosus. Transcatheter recanalization of the ductus re-established right pulmonary artery flow and growth. Ultimately, this allowed for complete surgical repair and restoration of normal perfusion of the right lung, leading to complete functional rehabilitation.

Keywords: Pulmonary arteries • Lung • Benign or congenital lesions • Cardiac catheterization/intervention

INTRODUCTION

Isolated, unilateral lung hypoplasia is a rare finding in paediatric patients [1]. Patients can be asymptomatic and present only in later life [2, 3] as the contralateral lung will make up for the functional loss in an attempt to maintain ventilation in keeping with perfusion. Unilateral lung hypoplasia is more commonly associated with conditions such as congenital diaphragmatic hernia that directly affect lung development by acting as space-occupying lesions. Another entity of abnormal vascular supply to one lung is that of cases with an aberrant origin of one pulmonary artery from the aorta—so-called hemitruncus arteriosus [4, 5]. These patients will present in cardiac failure with unrestricted blood flow to the anomalously perfused lung.

We argued that unilateral lung arterial blood supply, in the presence of contra-lateral normal parenchyma, bronchial tree and pulmonary venous return requires the committed search for the missing pulmonary arterial supply and, ideally, transcatheter rehabilitation. Flow to the disconnected pulmonary artery would promote growth to allow for later surgical reconnection with delivery of systemic venous blood to the non-perfused lung.

MATERIALS AND METHODS

A 10-year old girl was investigated on a general paediatric assessment unit for a chest infection. The chest X-ray revealed a hypoplastic right lung and scoliosis. Further history revealed that she was tiring more quickly than her peers and required more frequent rests during physical activity. Subsequently, CT chest (Fig. 1) showed a hypoplastic right lung, but the parenchyma was reported to be normal. There was a large left pulmonary artery (Fig. 1A), but no right pulmonary artery was identified. Both the left and right pulmonary veins were identified and were normally connected to the left atrium (Fig. 1B). The bronchial tree had a normal branching pattern bilaterally (Fig. 1C).

She was referred for cardiac assessment to rule out associated cardiac lesions and estimate right ventricular pressure. Cardiovascular examination at this time was normal, apart from the cardiac apex being shifted slightly to the right. There was a small patent foramen ovale, but otherwise the intracardiac anatomy was normal. There was no significant right ventricular hypertension. In the context of the chest CT having shown a normal right lung parenchyma, a normal bronchial tree and normal pulmonary veins, a decision was made to undertake cardiac catheterization. The plan was to search for a remnant of the right pulmonary artery and, if present, attempt catheter recruitment. This would ultimately allow for later surgical reconnection to the main pulmonary artery.

At catheterization, the CT finding of single left pulmonary artery with normal peripheral distribution and venous drainage was confirmed. Left pulmonary artery pressures were not significantly elevated. The aortogram showed a normal left-sided aortic arch with a blind ending ductal ampulla arising from the underside of the right-sided innominate artery (Fig. 2A). This suggested a spontaneously closed right-sided ductus arteriosus. Right pulmonary vein wedge contrast injections were possible through the patent foramen ovale. These demonstrated a hypoplastic right pulmonary arterial system (3 mm) with a normal branching pattern (Fig. 2B). These angiographic findings confirmed an anomalous origin of the right pulmonary artery from the occluded right-sided ductus. A 4 French right Judkins catheter was placed in the occluded ductal ampulla and the right-sided ductus arteriosus was recrossed with a 0.018 in. straight Terumo guidewire (Terumo Medical Corporation, Somerset, NJ, USA) into the disconnected right pulmonary artery. The Judkins catheter could not be advanced through the ductus. Therefore,
the Judkins catheter was exchanged for a Turbotracker 18 micro-perfusion catheter (Boston Scientific, Natick, MA, USA). This was successfully advanced over the Terumo wire into the right pulmonary arterial system. Test injections confirmed the position within the hypoplastic right pulmonary artery. The Terumo wire was then replaced with a 0.014-in. Thruway coronary wire (Boston Scientific, Natick, MA, USA). The ductus was then initially ballooned with a 5-mm Maverick coronary balloon (Boston Scientific) and then stented using a 5 × 24 mm Liberte Monorail coronary stent (Boston Scientific). Subsequent contrast injections showed that there was a mild proximal stenosis where part of the ductus had not been covered by the initial stent. A second Liberte stent (5 × 12 mm) was placed to cover this area (Fig. 2C). The procedure was uneventful and the patient was discharged home the following day on aspirin and dipyridamole to maintain the patency of the stents.

RESULTS

At outpatient review 2 months following catheterization, there was no significant change in the patient’s exercise tolerance. On echocardiography, the right pulmonary artery had increased in size to 8 mm. Diagnostic catheterization performed ~6 months after the initial procedure showed that the right pulmonary artery system had grown significantly with good distal arborization (Fig. 2D). The main right pulmonary artery, initially measuring 3 mm in diameter, now measured 12 mm compared with a normal expected pulmonary artery size of 13 mm. Mean right pulmonary artery pressure was 28 mmHg. The patient was referred for surgical repair.

Surgical findings were of the normal origin of the main and left pulmonary arteries with the complete absence of an intra-pericardial right pulmonary artery. The right pulmonary artery arose from a duct on the innominate artery and opened out behind the superior caval vein to a good-sized vessel with normal arborization. The ductus and right pulmonary artery were mobilized before establishing cardiopulmonary bypass. The ductus was doubly ligated and transected and all the remnants of the stent were removed. The opening was extended into the lower lobe of the right pulmonary artery to create a wide opening and a 16-mm Goretex conduit was anastomosed here. A large opening was made in the rightward side of the main pulmonary artery to receive the proximal conduit. After full rewarming, cardiopulmonary bypass was weaned on no inotropic support. The right atrial pressure was 6 mmHg and bypass time was 44 min.

The postoperative recovery was uneventful, and she was discharged on Day 5 postoperatively on aspirin. At 6-month post-operative follow-up, she had normal exercise capacity. Right ventricular systolic pressures estimated with cardiac ultrasound were normal. The previously noted scoliosis has completely resolved and she has been discharged from an orthopaedic follow-up.

DISCUSSION

Isolated unilateral lung hypoplasia in the setting of a normal bronchial tree, normal pulmonary venous drainage and absent pulmonary arterial supply, should prompt a committed search for a native pulmonary artery system to the hypoplastic lung. Catheter recanalization should be aggressively attempted to try to rehabilitate perfusion and growth of the disconnected pulmonary arterial system. Re-establishing blood supply promotes growth of the native pulmonary artery, without conveying a functional benefit as pulmonary venous blood is delivered to the disconnected pulmonary artery. The growth of the pulmonary arterial tree allows ultimately for surgical reconnection to the main pulmonary artery with delivery of systemic venous blood to the lung. This re-establishes normal ventilation-perfusion, achieving complete functional rehabilitation of the chronically disconnected lung.

Conflict of interest: none declared.
REFERENCES


Figure 2: (A) Aortogram showing left-sided arch with blind ending ductal ampulla arising from the innominate artery. (B) Retrograde pulmonary vein injection demonstrating disconnected, hypoplastic right pulmonary artery (RPA) system. (C) Angiogram after stent recanalization of the occluded ductus. (D) Repeat angiogram of RPA system 6 months after ductal stenting.