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A large, single pulmonary arteriovenous fistula presenting hours after birth

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MeSH: Heart Defects, Congenital/*diagnosis/*surgery, Infant, Newborn, Pulmonary Artery/*abnormalities

Introduction

We report a case of a single, large pulmonary artery to left atrial fistula presenting within hours of birth. Symptomatic fistulas of this type are exceptionally rare in the neonatal period. We include images of the fistula obtained during echocardiography and cardiac catheterisation. This case highlights the importance of intra-operative echocardigraphic guidance during surgical ligation of fistulas of this type.

Case report

A term baby was born in a district general hospital following a normal antenatal course. She was in good condition at birth, but six hours following delivery was noted to be cyanotic, with capillary oxygen saturations of 80% in room air and a poor response to supplementary oxygen. A prostaglandin E2 infusion was commenced and she was transferred to the regional centre.

Following transfer it was noted that the prostaglandin infusion had made no significant difference to the oxygen saturations. There was a palpable left ventricular impulse, a grade 2/6 continuous murmur loudest below the right clavicle and 2cm of palpable hepar. Femoral pulses were easily felt.

An electrocardiogram showed sinus rhythm with prominent left ventricular voltages. A plain film of chest indicated moderate cardiomegaly with prominent vascularity at the right hilum and relative oligaemia of the peripheral lung fields (Figure 1).

The findings on echocardiography were highly suspicious of a large fistula between a major branch of the right pulmonary artery and lower lobe vein (Figure 2).

Figure 1 The chest X-ray following intubation shows cardiomegaly and some asymmetry of pulmonary vascularity.

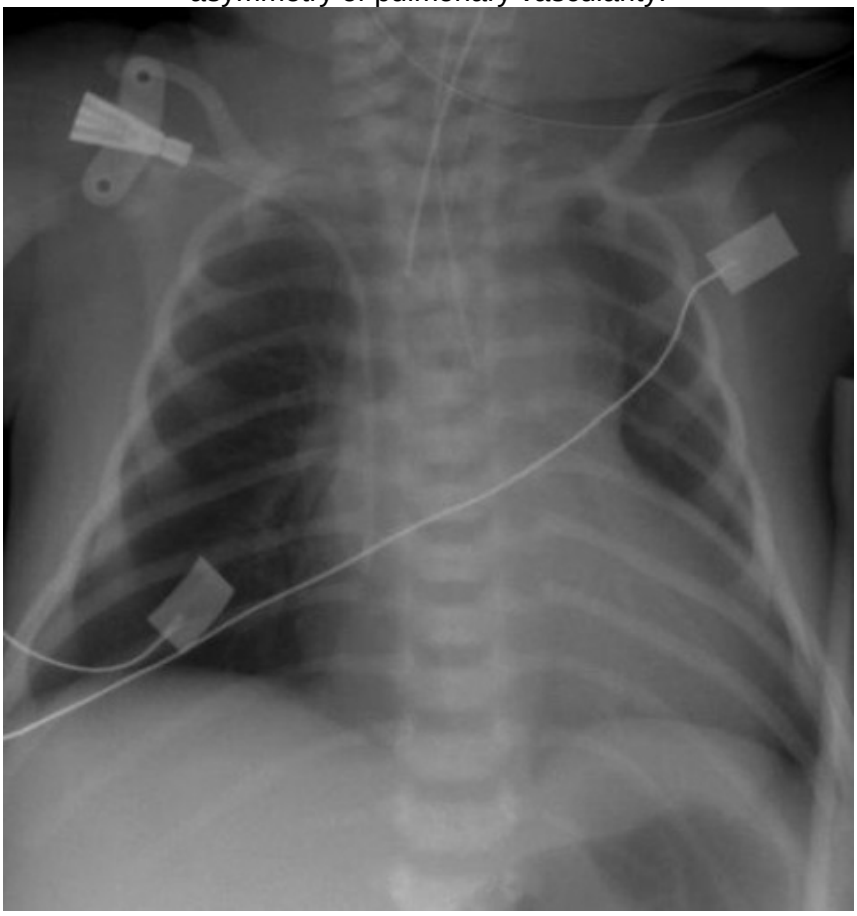
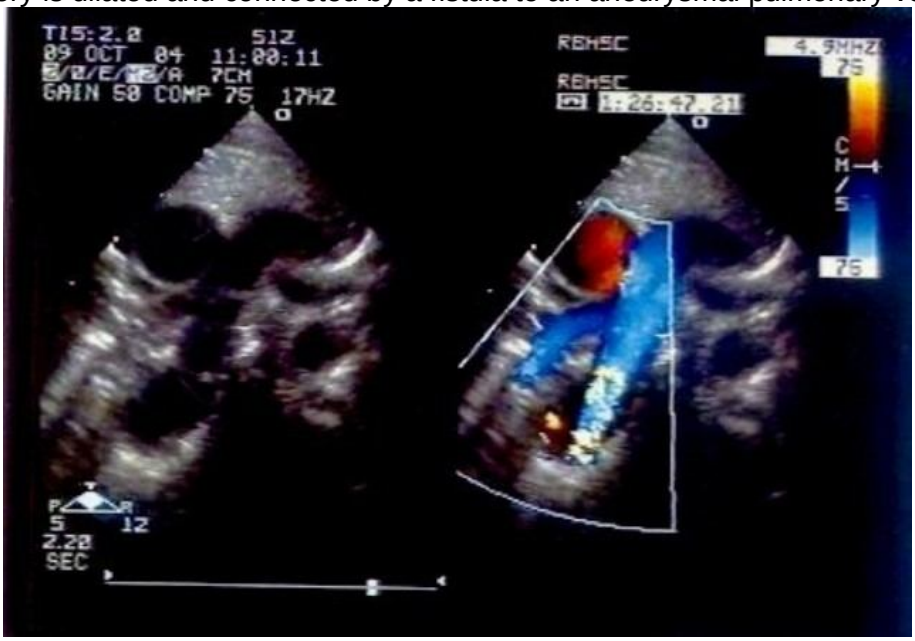


Figure 2 The echocardiogram (with colour comparison) shows the right pulmonary artery is dilated and connected by a fistula to an aneurysmal pulmonary vein.



The venous portion of the malformation was aneurysmal and communicated freely with the left atrium. There was torrential flow through the fistula into the left atrium. Bubble echocardiography also supported the diagnosis of a large right-to-left shunt.
The prostaglandin infusion was stopped and the patient was treated with diuretics. Her clinical course deteriorated over the next two days, with increasing cyanosis, worsening cardiac failure and frequent apnoea. The patient was transferred to intensive care, where she was treated with inotropes, high frequency oscillatory ventilation and inhaled nitric oxide. This was in an effort to reduce resistance in the normal segments of the pulmonary artery tree.
The patient's condition remained extremely unstable and surgery was carried out on day three of life. The procedure was carried out without intraoperative echo guidance via a right thoracotomy. The operation was technically difficult because of poor oxygenation and consequent hyperventilation hampering lung dissection. The abnormal vessel entered the dilated left atrium posterior to the hilum of the lung, and was not identified at this procedure. Tissue thought to contain the vessel was ligated, following which there was a dramatic improvement in systemic oxygenation, which was taken as evidence of occlusion. In fact this was probably only caused by temporary distortion and kinking of the fistulous vessel.
Post operatively, the improvement in oxygen saturation proved transient and the patient's condition again became very unstable. The echocardiographic findings were unchanged and it was concluded that the target vessel had not been successfully ligated.
Cardiac catheterisation was performed to further delineate the anatomy (Figures 3,4).

Figure 3 The PA angiogram shows a catheter passed through the fistula via the right pulmonary artery. The pulmonary venous aneurysm communicates freely with the left atrium.

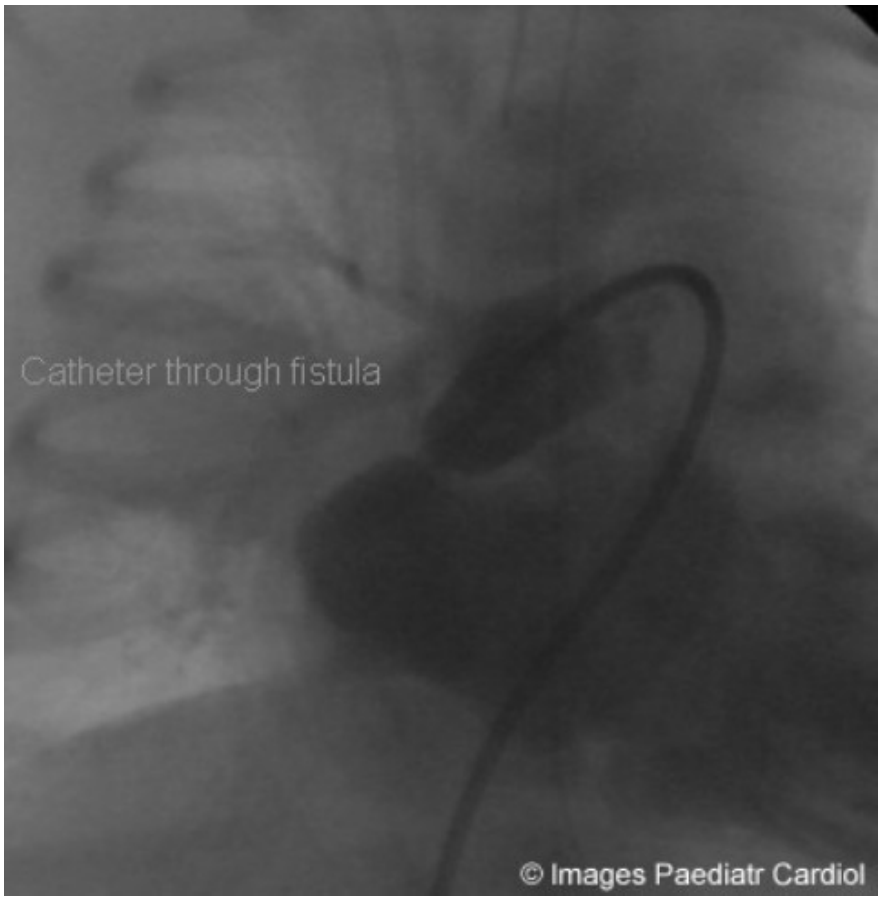
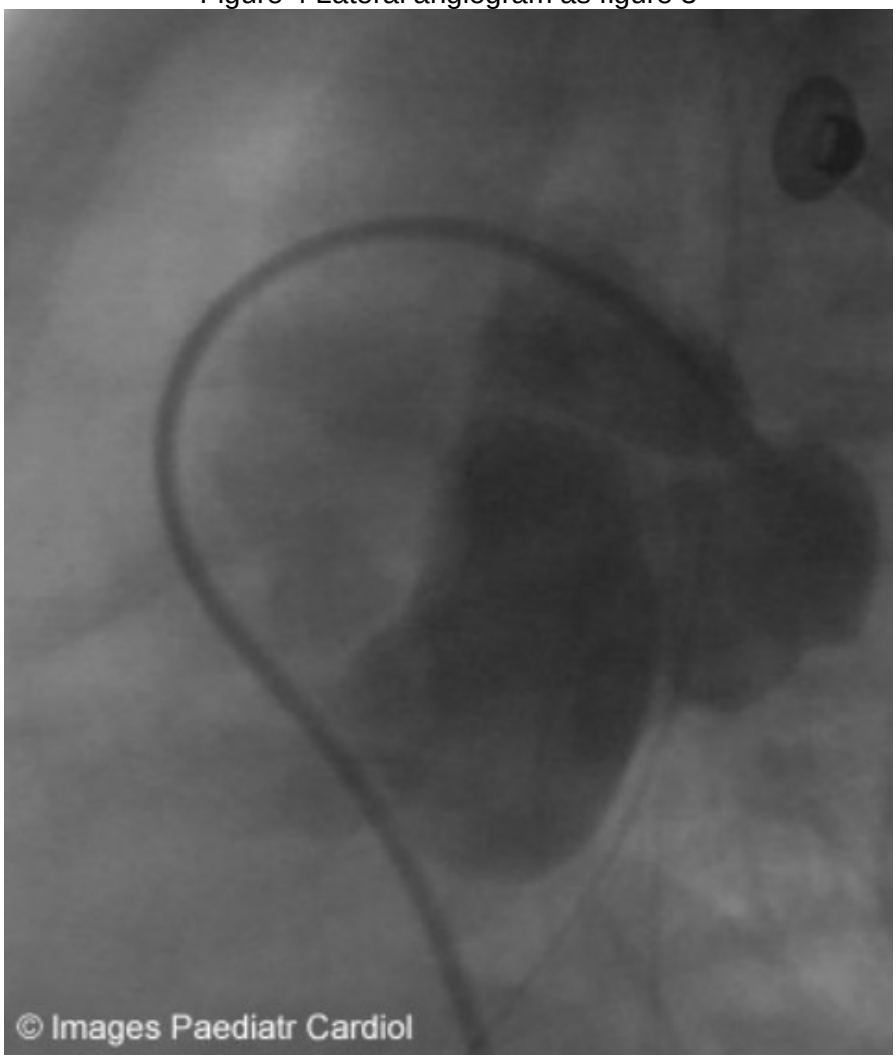


Figure 4 Lateral angiogram as figure 3



This clearly demonstrated a large fistulous connection, measuring around 4 millimetres. The fistula was temporarily occluded with a balloon catheter (Figures 5,6), resulting in a rise in oxygen saturations from 60% to 99%.

Figure 5 The PA and lateral angiogram shows a balloon catheter partially occluding the fistula, inducing a rise in systemic oxygen saturation.

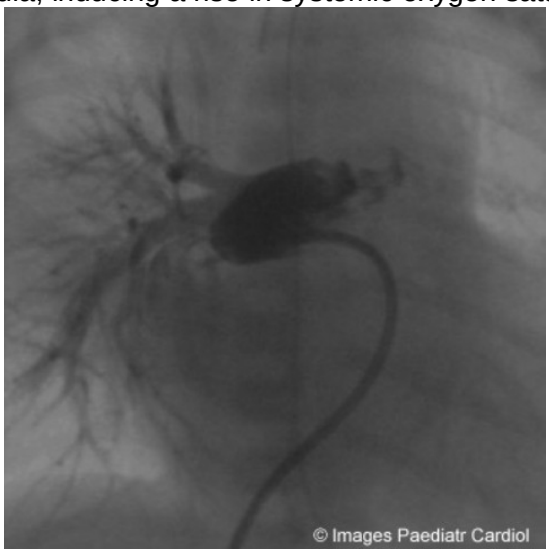
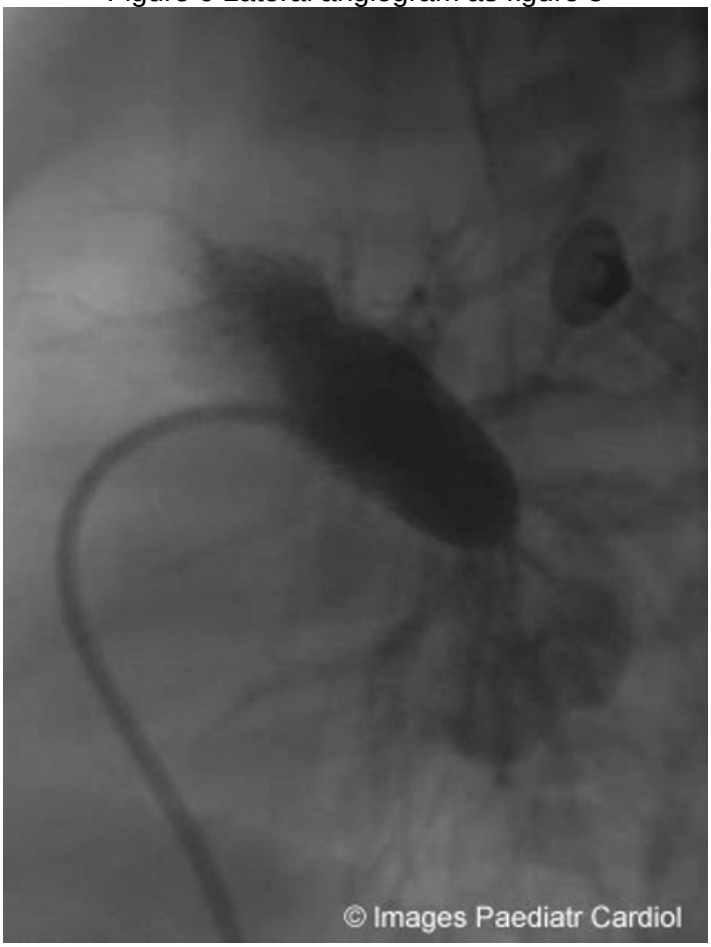


Figure 6 Lateral angiogram as figure 5



An attempt was made to place an 8mm Cook detachable coil (Cook Inc, Bloomington, IN) in the feeding vessel. However it was impossible to achieve a satisfactory position and no device was deployed.

The following day the child underwent further surgery. During this procedure trans-thoracic echocardiography was used for guidance. The distal portion of the abnormal vessel was found closely applied to, and virtually indistinguishable from the wall of the left atrium. The vessel was ligated with an immediate rise in oxygen saturation and blood pressure. Closure of the fistula was confirmed by echocardiography.

Post-operatively there was a prolonged recovery period, complicated by a right phrenic nerve palsy, poor weight gain and lower respiratory tract infection. The patient was discharged from hospital on no medications at two months of age. She is currently acyanotic and thriving with no respiratory symptoms.

Discussion

Symptomatic pulmonary arteriovenous malformations are rarely encountered in the neonatal period. There have been relatively few cases of pulmonary artery to left atrial fistula reported; most of which have presented in the third decade of life.[1] In a recent case series 15.8% presented with the severe neonatal form of this condition.[2] A previous case series indicated that the mortality in symptomatic pulmonary arteriovenous malformations in those under six months of age is 43%.[3]

The treatment options for pulmonary arteriovenous malformations are transcatheter occlusion or surgical ligation. Current opinion favours transcatheter occlusion of

these lesions if possible.[4,5] Previously it was thought that surgical resection was the optimal mode of therapy. Unfortunately in our case the fistula was too large for coil occlusion. In a larger patient a reasonable alternative would be use of an Amplatzer occlusion device, e.g. duct occluder or muscular VSD device (AGA Medical Corporation, MN).[6,7]

A learning point from this case is the importance of intra-operative echocardiolographic guidance. Echo guidance at the second procedure allowed the surgeon to test the effect of occluding the presumed target vessel before complete ligation.

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