

Anomalous origin of the left pulmonary artery

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A 4-year-old girl presented to our hospital with a diagnosis of a subpulmonic ventricular septal defect. It was reported that she had been markedly symptomatic from early infancy with failure to

thrive, recurrent lower respiratory tract infection and interruption of breast feeding. She was on furosemide 20mg and spironolactone 6.25mg orally once a day.

On physical examination her weight was 14.5kg and her height was 93cm. Her blood pressure was 104/51mmHg in the left arm. No cyanosis was noted. Her chest was clear with good air entry. Her precordium was active and bulging with the apical impulse displaced downward and laterally. She had a pansystolic murmur of 3/6 best heard over the left lower sternal border. The liver was palpable 3cm below the right costal margin. She had a trace of pedal edema.

Her hematocrit was 37.6%. Other hematologic profiles were also within normal limits. The electrocardiogram (EKG) showed sinus rhythm at 108bpm and no remarkable abnormality was noted. Chest X-rays showed moderately increased cardiothoracic ratio, with marked left lung plethora. Echocardiography showed a dilated left atrium, left ventricle and main pulmonary artery; large subpulmonic VSD of 11mm with left to right shunt at a pressure gradient of 50mmHg. Left ventricular systolic function was good.



FIGURE 1: Pulmonary artery angiography showed an absent LPA

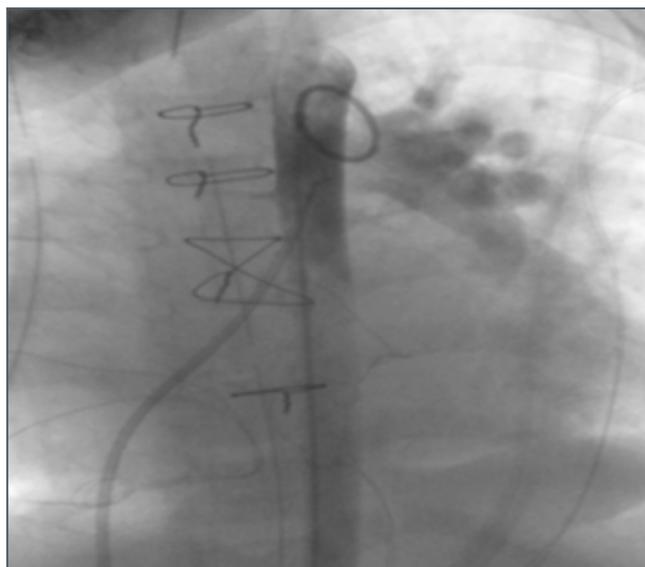


FIGURE 2: Aortic injection showed the left pulmonary artery coming off the descending aorta

There was moderate mitral regurgitation noted.

The patient was scheduled for surgery. After the aorta and the caval veins were cannulated, the surgeon noticed that massive pulmonary venous return was still coming to the left atrium. External anatomy revealed an absent left pulmonary artery (LPA). Intra-operative transesophageal and epicardial echocardiography failed to show the origin of the LPA. A day after the surgery, the patient was taken to the catheterisation laboratory and cardiac angiography was performed. The mean pulmonary artery pressure was 25mmHg and the mean aortic pressure was 61mmHg. Pulmonary artery angiography showed an absent LPA (Figure 1). Aortic injection showed the left pulmonary artery coming off the descending aorta (Figure 2). The patient was directly transferred to the operating theatre and the chest was reopened. The LPA was noted to be coming off the descending aorta. Under cardiopulmonary bypass (CPB), the LPA was transected from the aorta and the aorta was over sewn. The LPA was directly attached to the MPA. CPB was discontinued. The serum potassium was noted to be 7.8mEq/L. CPB was re-established and filtration done over 4 hours and 4 liters of fluid removed. In spite of this, the serum potassium remained high. A peritoneal dialysis catheter was inserted and peritoneal dialysis started. The patient was then transferred to the ICU in poor condition and died a few hours later.

Anomalous origin of either of the pulmonary arteries is a relatively rare congenital anomaly.⁽¹⁾ Anomalous origin of the right pulmonary artery has been more commonly reported.^(1,2) An anomalous origin of the left pulmonary artery (LPA) from the ascending aorta is very rare. Anomalous origin of the left branch pulmonary artery usually occurs in association with Tetralogy of Fallot.⁽³⁻⁵⁾ Walter, et al. reported a case of the LPA coming off the ascending aorta, in association with a right sided aortic arch and right patent ductus arteriosus.⁽⁶⁾ Our patient presented with a rare occurrence of LPA coming off the descending aorta in association with sub-pulmonic VSD.

Unfortunately, our patient did not survive the surgery. Careful attention to the asymmetric lung perfusion on frontal chest X-rays

should have led to pre-operative cardiac catheterisation and an optimal outcome.

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