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Brief Report

Rapid onset of intrapulmonary arteriovenous shunting after surgical repair of tetralogy of Fallot with pulmonary atresia

Victor D. Ofoe, Usha Pratap, Zdenek Slavik

Royal Brompton and Harefield NHS Trust, Paediatric Surgical Unit, Harefield Hospital, Middlesex, UK.

Abstract We describe a 2-year-old girl with tetralogy of Fallot and pulmonary atresia, palliated as a neonate with a right modified Blalock Taussig shunt, who developed severe cyanosis following total correction in the absence of corresponding evidence of parenchymal lung disease on the chest X-ray. Selective pulmonary angiography showed new intrapulmonary shunting involving only the right middle and lower lobes only. The cyanosis resolved rapidly subsequent to inhalation of nitric oxide. To our knowledge, this is the first documented case of rapid onset of localised intrapulmonary right-to-left shunting, involving only two lung lobes, following biventricular repair for complex congenital heart disease.

Keywords: cyanosis; complex congenital heart disease; intrapulmonary arteriovenous shunting

THE MOST FREQUENT CAUSES OF PROFOUND cyanosis following surgical repair of congenital cardiac malformations are pulmonary hypertensive crises and residual right-to-left intracardiac shunts. A more gradual onset of cyanosis also occurs after construction of a superior cavopulmonary anastomosis.[1] We report here an unusual case of severe cyanosis, shown to be due to intrapulmonary right-to-left shunting, which occurred 20 hours after an otherwise successful biventricular repair of tetralogy of Fallot with pulmonary atresia.

Case Report

The patient was a 2-year-old girl with tetralogy of Fallot and pulmonary atresia who, in the neonatal period, had been palliated by construction of a right modified Blalock-Taussig shunt. She subsequently underwent total repair, with ligation of the Blalock-Taussig shunt, reconstruction of the right ventricular outflow tract by insertion of a pulmonary homograft, and patch closure of the ventricular septal defect. She remained haemodynamically stable and fully saturated while ventilated. Extubation was carried out successfully 18 hours postoperatively, but two hours later, she became progressively desaturated, with oxygen falling to below 80%, with no response when concentrations of oxygen were increased by ventilation through a face mask. Her chest X-ray at this stage showed minimal bilateral infiltrates, which were not compatible with the degree of desaturation (Fig. 1). She was reintubated when her saturations reached 70%. A contrast echocardiogram showed no intracardiac right-to-left shunting, and cardiac catheterisation demonstrated a small residual ventricular septal defect with a shunt from left-to-right. There was no obstruction within the right ventricular outflow tract, nor within the pulmonary arteries. Right ventricular systolic pressure was 52 mmHg, with a simultaneous right femoral arterial systolic pressure of 80 mmHg. The ratio of mean pulmonary arterial and aortic pressures was 2 to 3, with the mean pulmonary arterial pressure measured at 34 mmHg. Selective pulmonary arteriography showed transit of contast through the right middle and lower lobes in less than one and a half cardiac cycles, with a heart rate of 160 per minute (Fig. 2a), demonstrating significant intrapulmonary arteriovenous shunting. The remaining parts of the right and the left lung showed a normal pattern of perfusion without any

Correspondance to: Dr Z Slavik, Royal Brompton and Harefield NHS Trust, Paediatric Surgical Unit, Harefield Hospital, Hill End Road, Harefield, Middlesex, UK. Tel: 01895 828554; Fax: 01895 828554; e-mail: slavik@ hotmail.com

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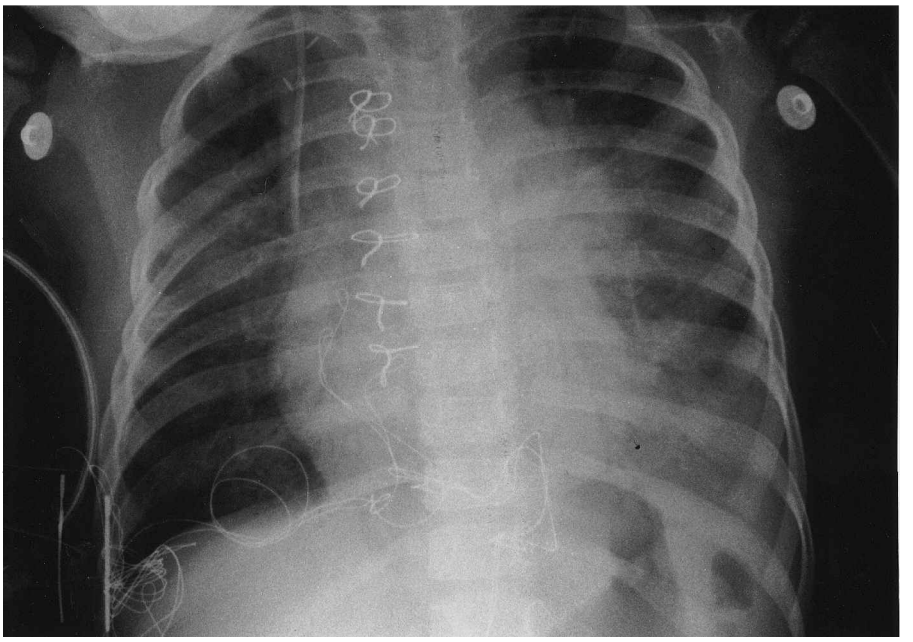

Figure 1.
The plain chest radiograph following extubation when the patient was acutely desaturated.

residual findings. No evidence of intrapulmonary right-to-left shunting was found on retrospective review of her preoperative angiograms (Fig. 2b). She was then treated with inhaled nitric oxide, given at 5 parts per million, for four days, with resolution of her cyanosis and restoration of normal oxygen saturations. Her further post-operative course was complicated by recurrent intrabronchial bleeding. She was gradually weaned from the ventilator and extubated on the 26th postoperative day. She was discharged home on the 46th postoperative day when saturations of oxygen were normal while breathing air.

Discussion

Intrapulmonary arteriovenous shunting is an inevitable complication following the bidirectional cavopulmonary anastomosis.[1,2] This is followed by development of macroscopic pulmonary arteriovenous malformations, and progressive cyanosis in some of the patients. Such malformations have been documented as early as 72 hours after construction of a bidirectional cavopulmonary anastomosis.[3] The reason for such right-to-left shunting in this setting has been ascribed to the absence of pulsatile flow in the pulmonary arteries, and/or the diversion of hepatic venous flow away from the pulmonary circulation.[1,4]

Intrapulmonary right-to-left shunting has also been described in patients with systemic disorders, such as Osler-Weber-Rendu syndrome or liver disease, the so-called hepatopulmonary syndrome.[5] The onset of cyanosis in these cases is usually gradual, and these patients share the paucity of hepatic venous effluent reaching the pulmonary circulation with the patients who have undergone construction of a bidirectional cavopulmonary anastomosis.

Young infants and children have been shown to have primordial arteriovenous connections in their lungs.[6] These connections may reopen acutely, leading to the development of clinically significant pulmonary arteriovenous shunting. Physiological studies have shown that these channels enlarge with increases in the pulmonary arterial systolic pressure.[7] The increase in flow of blood to the lungs in our patient following her corrective surgery may have resulted in the reopening of these primordial intrapulmonary channels. The site of the shunt could be related to regional distribution of blood flow in the lungs, as there was a mild stenosis to the branch of right pulmonary artery supplying the right upper lobe, which was not involved in the right-to-left.

Acute respiratory distress syndrome is another well-recognised cause of hypoxaemia following cardiac surgery.[8] It is difficult, however, to explain the degree of progressive hypoxaemia encountered in our patient on the basis of the acute respiratory distress syndrome, particularly since only two lobes of the right lung were affected, and there were no positive radiological findings.

Inhalation of nitric oxide led to rapid resolution of the cyanosis in our patient. This agent has also been used effectively in the treatment of patients with pulmonary arteriovenous malformations following construction of a bidirectional cavo-

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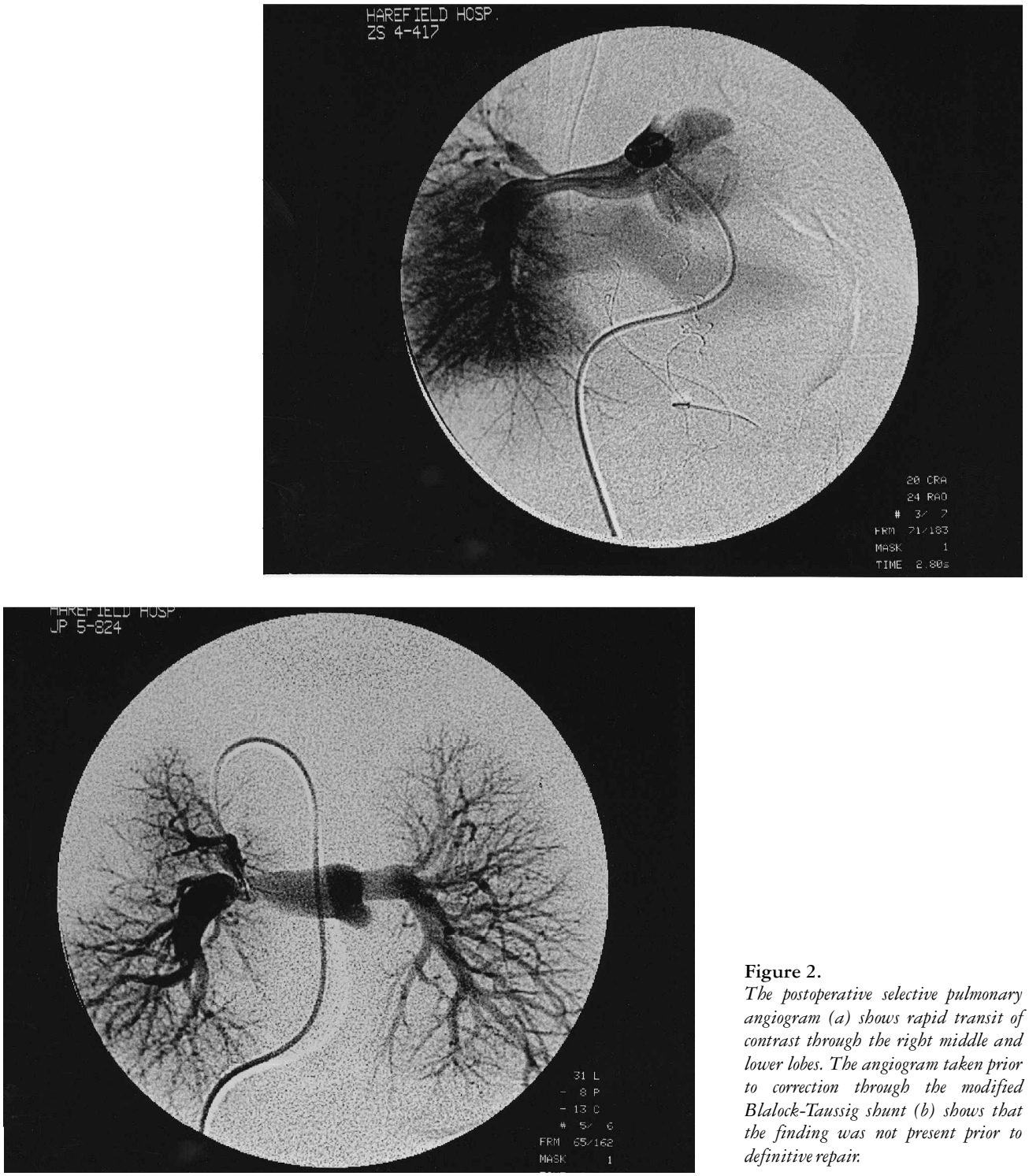

Figure 2.
The postoperative selective pulmonary angiogram (a) shows rapid transit of contrast through the right middle and lower lobes. The angiogram taken prior to correction through the modified Blalock-Taussig shunt (b) shows that the finding was not present prior to definitive repair.

pulmonary anastomosis.[2] We believe that the inhalation of nitric oxide redistributed the flow of blood away from the segments of the lung with intrapulmonary shunting, and thus improved the ventilation perfusion mismatch present in our patient.

To our knowledge, acute onset of clinically important intrapulmonary arteriovenous shunting, involving only two lobes of one lung, has never previously been reported following biventricular repair for complex congenital heart disease. The etiology is likely to be multifactorial, and further investigation is needed to find the risk factors involved in its development.

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