Cardiol Young 2003; 13: 571–573

ISSN 1047-9511

Brief Report

Haemoptysis in a patient with tetralogy of Fallot: a combined surgical and interventional approach

W. Budts,[1] P. Moons,[1] M. Gewillig[2]

[1]*Division of Congenital Cardiology,* [2]*Division of Paediatric Cardiology, University Hospitals Leuven, Belgium*

Abstract Haemoptysis may occur in patients with tetralogy of Fallot and major aorto-pulmonary collateral arteries. We describe such a patient in whom bleeding from a major aorto-pulmonary collateral artery produced severe pulmonary haemorrhage. Interventional closure of the artery could not be performed because it perfused the native pulmonary arteries. Instead, we inserted a conduit between the right ventricle and the native pulmonary arteries, followed by percutaneous closure of the collateral artery. Our patient demonstrates the increasing necessity for combined surgical and interventional procedures.

Keywords: Major aorto-pulmonary collateral artery; interventional cardiology; Amplatzer device; multidisciplinary

Some patients with tetralogy of Fallot present with severe pulmonary stenosis or pulmonary atresia. In this setting, pulmonary arterial supply can be via major aorto-pulmonary collateral arteries. It is then not uncommon for pulmonary haemorrhage to occur when it is not possible to perform corrective surgery. If the haemorrhage is life-threatening or to prevent recurrence, it is then feasible to embolise the culprit artery.

We were recently challenged by such a case of haemoptisis in this setting in which the culprit artery was also the only vessel that perfused the lower lobe of the left lung and the native pulmonary arteries. Occlusion of this vessel would have led to a massive pulmonary infarction and significant systemic desaturation. Only by performing a hybrid interventional and surgical procedure were we able to achieve an acceptable solution.

Case report

A 16-year-old girl with tetralogy of Fallot was referred to our tertiary care hospital after a third episode of severe haemoptysis in a period of three months. Tetralogy had been diagnosed immediately after birth, confirmed by echocardiography, and complicated by pulmonary atresia, the pulmonary arterial supply derived from major aorto-pulmonary collateral arteries. The systemic saturation was acceptable (85%), and the paediatric cardiologist planned to treat this patient conservatively for her first years of life. At the age of 5 years, the first angiogram was done to define the anatomy of the major aorto-pulmonary collateral arteries, and the development of the native intrapericardial pulmonary arteries (Fig. 1). Because the risk for repair was considered to be too high, it was decided not to change the conservative approach to treatment. The plan was to schedule palliative surgery, such as unifocalization of the arteries supplying the left lung and construction of a Blalock-Taussig shunt, subsequent to any marked decrease in systemic saturation. Four months prior to her admission to hospital because of a third episode of haemoptysis, the major aorto-pulmonary collateral artery which perfused the left upper lung, and which was known to have a pressure of 18 over 11 mmHg, with a mean of 14 mm, had been dilated and stented. The systemic saturation had then improved from 82% to 87%.

At the first two episodes of haemoptysis, we had not been able to localise the origin of bleeding using bronchoscopy and aortography, and therefore we continued with conservative treatment. No triggers for bleeding, such as an infection of the respiratory tract infection or coagulative disorders, could be identified.

Correspondence to: Werner Budts, Division of Cardiology, Congenital Cardiology, Herestraat 49, B-3000 Leuven, Belgium. Tel: 32 16 344235; Fax: 32 16 344240; E-mail: Werner.Budts@uz.kuleuven.ac.be

Accepted for publication 30 May 2003

At the current hospital admission, the cardiopulmonary circulation was hyperdynamic, with raised blood pressure and increased pulse. We deemed this hyperdynamic state to be triggered by anxiety, because blood pressure and pulse had previously always been within normal ranges. The hematocrit was now only 50%, 5% lower when compared with previous blood tests. Since stenting the collateral artery, systemic saturation had been maintained at 87%, yet the patient was suffering from progressive asphyxia, indicated by a saturation of 78% on admission, with a subsequent decrease to 67% shortly after hospitalisation. Intubation, mechanical ventilation, and an urgent bronchoscopy were necessary to extract thrombus from the airways. Although the patient was sedated deeply, and the perfusion pressures decreased to normal levels, active bleeding was identified in the left lower lung. It was estimated that the patient lost a volume of 500 ml of blood. Based on the bronchoscopic data, and the fact that the major aorto-pulmonary collateral artery, which perfused the left lower lung, was known to be perfused at high pressure, we considered it likely that this collateral artery was responsible for the recurrent episodes of haemoptysis. Closure was suggested, but we knew such a procedure would exacerbate the poor condition, since this artery also perfused the native pulmonary arteries via a stenotic side branch (Fig. 1). We opted instead for a combined surgical and interventional approach.

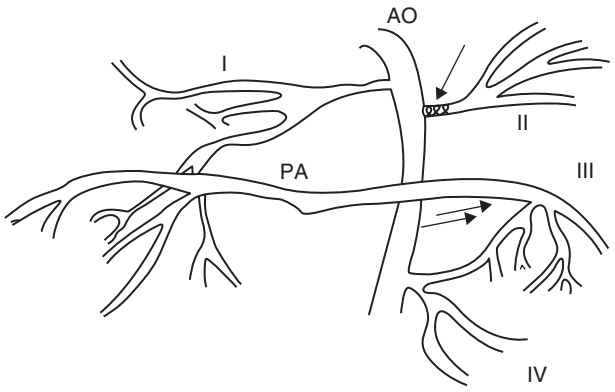

Figure 1.
Anatomy of the major aorto-pulmonary collateral arteries. The collateral arteries are numbered from I to IV. The first artery perfuses the right upper and lower lung at low pressure. The second artery perfuses the left upper lung through a stented orifice (single arrow), at high pressure. The third artery perfuses the left lower lung at high pressure and, via a stenotic side branch (double arrow), supplies also the native pulmonary arteries at low pressure. The final collateral artery perfuses the left lower lung at low pressure. AO: descending aorta; PA: native pulmonary arteries.

First, using cardiopulmonary bypass, the surgeon inserted a 10 mm Gore-Tex graft between the right ventricle and the native pulmonary arteries (Fig. 2a). Two weeks later, the collateral artery was closed percutaneously with a 12 mm Amplatzer occluder (Fig. 2b). No haemoptysis occurred either after surgery or before intervention. Prior to insertion of the conduit, systolic pressure in the native pulmonary artery was 20 mmHg, which rose to 62 mm after insertion of the conduit. After occlusion, the systolic pressure in the pulmonary arteries decreased to 55 mmHg, with the distal branches of the collateral now perfused via the native pulmonary arteries. This resulted in a

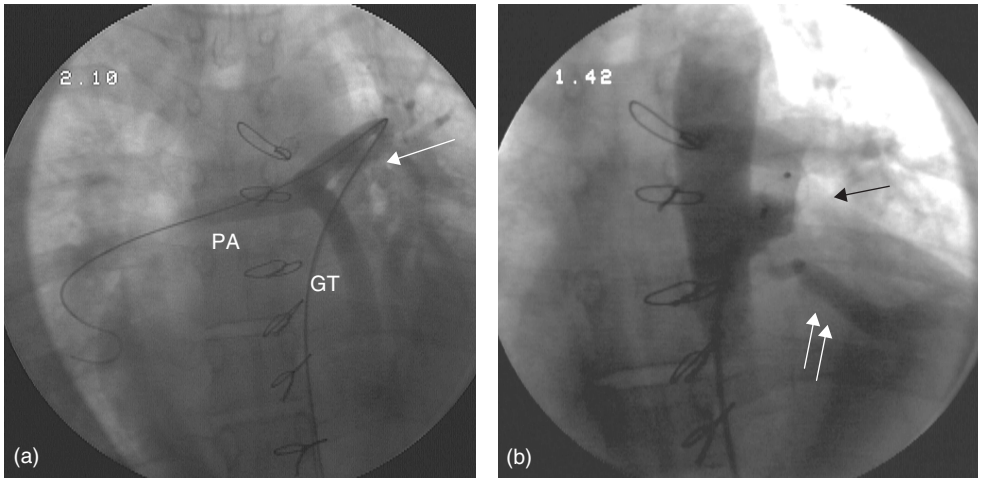

Figure 2.
An angiogram (Fig. 2a) reveals the anatomy of the native pulmonary arteries. A guide wire is positioned in the native pulmonary artery via the third major aorto-pulmonary collateral artery as shown in Figure 1 (arrow). Contrast fills the native pulmonary arteries, showing also the 10 mm Gore-Tex graft. PA: pulmonary arteries; GT: Gore-Tex. An aortogram (Fig. 2b) confirms the position of the 12 mm Amplatzer occluder (single arrow) distal to the mouth of the fourth major aorto-pulmonary collateral artery shown in Figure 1 (double arrow). Minimal flow of contrast to the native pulmonary arteries was seen just after closure of the collateral artery.

https://doi.org/10.1017/S1047951103001203 Published online by Cambridge University Press

reduction by half in systolic pressure, and an increase in systemic saturation to more than 85%. The patient recovered well and has remained, 12 months after the procedures, free of episodes of haemoptysis. The plan for subsequent treatment will depend on the clinical and haemodynamic evolution. Should definitive repair not be possible, heart–lung transplantation will be unavoidable.

Discussion

Tetralogy of Fallot is one of the most common congenital heart defects followed-up in paediatric and adult clinics for congenital cardiac disease. Most of the patients with tetralogy now undergo repair at young age, and long-term follow-up is needed for early detection of right ventricular dysfunction caused by pulmonary regurgitation, or arrhythmias.[1]

A small proportion of patients with tetralogy, however, present with severe pulmonary stenosis or pulmonary atresia, with major aorto-pulmonary collateral arteries feeding the pulmonary circulation.[2] Many of these patients can never undergo "corrective" surgery. They are frequently confronted with progressive cyanosis, and develop secondary erythrocytosis. Palliative treatment is now able to improve the outcome for these patients, and often provides a bridge to heart-lung transplantation.[3] Massive and life-threatening haemoptysis may occur in such patients, nonetheless, usually originating from the aneurismal dilation of hypertrophied bronchial vessels, the erosion of varicose bronchial vessels into the airway, or rupture of the major aorto-pulmonary collateral arteries.[4] Such pulmonary haemorrhage is frequently triggered by infections of the respiratory tract, and co-existing clotting disorders, especially in patients with secondary erythrocytosis. If the bleeding compromises the cardiopulmonary haemodynamics, embolisation of the culprit artery is indicated.[5–7] Asphyxia, which is caused by the clotting of blood in the airways, is uncommon, but may result in life-threatening ventilatory impairment.[8] This complication needs management using flexible or rigid bronchoscopes.[9]

Such bronchoscopic extraction of blood clots was necessary in our patient, and it was suggested that the bleeding originated from major aorto-pulmonary collateral artery, because active bleeding could be localized in the left lower lung, the territory of blood supply of this collateral. Instant embolisation of this vessel was not possible since a stenotic side branch of the collateral artery also perfused the native pulmonary arteries. Embolisation of the artery, therefore, would likely have resulted in loss of the native pulmonary arteries. Hence, our therapeutic goal was to lower the pressure in the distal part of collateral artery, and we requested a multidisciplinary approach involving cardiac surgeons, paediatric cardiologists, and adult cardiologists with expertise in congenital cardiac disease. The first step was the insertion of a conduit between the right ventricle and the native pulmonary arteries. The second step was the percutaneous closure of the collateral using an Amplatzer occluder. These procedures achieved our major goal, namely to produce lower pressures in the distal part of the territories supplied by the major aorto-pulmonary collateral artery. In the first 12 months after the procedures, there have been no recurrent episodes of haemoptysis. Our experience illustrates that collaboration is now critical in caring for patients with congenital cardiac disease. The combined approach offers substantial advantages. Isolated surgery, or a simple percutaneous intervention, would not have helped our patient.

Acknowledgement

We thank Prof Wim Daenen, from our Division of Congenital Cardiac Surgery, with whom the different options for treatment were discussed, and who performed the surgical intervention.

References

1. Moodie D. Diagnosis and management of congenital heart disease in the adult. Cardiol Rev 2001; 9: 276–281.
2. Warnes C. Tetralogy of Fallot and pulmonary atresia/ventricular septal defect. Cardiol Clin 1993; 11: 643–650.
3. Pigula F, Gandhi S, Ristich J, et al. Cardiopulmonary transplantation for congenital heart disease in the adult. J Heart Lung Transplant 2001; 20: 297–303.
4. Haroutunian L, Neill C. Pulmonary complications of congenital heart disease: hemoptysis. Am Heart J 1972; 84: 540–559.
5. Suzuki Y, Fukuda I, Kohno M, Jikuya T, Wada M, Tsutsui T. Preoperative therapeutic embolization for massive hemoptysis from bronchial collaterals in tetralogy of Fallot – a case report successfully treated by total correction. Nippon Kyobu Geka Gakkai Zasshi 1990; 38: 472–476.
6. van der Weijden P, Baur LH, Kool LJ, Vliegen HW, van der Wall EE. Embolization as a treatment of life-threatening haemoptysis in an adult with tetralogy of Fallot with pulmonary atresia. Int J Card Imaging 1998; 14: 123–126.
7. Gonzalez J, Ruiperez JA, Garcia Almagro FJ, Garcia Medina J, Capel A, Valdes M. Tetralogy of Fallot complicated by severe hemoptysis in the adult. Rev Esp Cardiol 2001; 54: 1002–1004.
8. Ooe K, Nakada H, Udagawa H, Shimizu Y. Severe pulmonary hemorrhage in patients with serious group A streptococcal infections: report of two cases. Clin Infect Dis 1999; 28: 1317–1319.
9. Arney K, Judson M, Sahn S. Airway obstruction arising from blood clot: three reports and a review of the literature. Chest 1999; 11: 293–300.