Tetralogy of Fallot with critical biventricular dysfunction: is surgical correction achievable?

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Abstract

We describe the case of a 32-month-old patient from a developing country with tetralogy of Fallot associated with a severe biventricular dysfunction. This association is rare but makes the surgical strategy complex and potentially contraindicated. An acute severe hypoxic episode led us to perform palliative rescue intervention involving the placement of an undersized systemic-to-pulmonary shunt. This surgery was well tolerated and allowed a fast and impressive recovery of the ventricular function, making complete repair possible.

Keywords: Tetralogy of Fallot • Biventricular dysfunction • Left ventricular dysfunction • Blalock–Taussig shunt

INTRODUCTION

Timing and indications of surgical techniques, systemic-to-pulmonary shunt or complete repair, remain controversial in patients with tetralogy of Fallot (TOF) [1]. In developing countries, most congenital heart diseases are not diagnosed until late in infancy. This is why patients with a late presentation of TOF are frequently encountered in surgical patients in a humanitarian setting [2]. Even in these late forms of TOF, the presence of a severe biventricular dysfunction is a rare finding, and makes the surgical strategy complex and potentially dangerous. In this situation, timing and indications are of particular importance.

CASE PRESENTATION

The patient came from Congo-Brazzaville and was referred to us by the French humanitarian association ‘la Chaine de l’Espoir’. On arrival in France, he was aged 2 years and 8 months, weighed 8.6 kg and was 82 cm in height. His general condition was precarious due to malnutrition. He was eupneic with 83% blood oxygen saturation in room air. Haemoglobin and haematocrit values were 17,5 g/dl and 63%, respectively. Echocardiography confirmed a regular form of TOF (Fig. 1) but showed a severe biventricular dysfunction with a left ventricular ejection fraction of 15%. This condition contraindicated an immediate surgical intervention.

The biventricular dysfunction was initially attributed to malnutrition, at least in part, and we decided to defer the surgery to improve his general condition. Unfortunately, after 1 month of nutritional rehabilitation and improvement in muscular tone, cardiac function remained very impaired. Echocardiographic examination of a left intraventricular thrombus suggested aggravation (Video 1).

Hydroelectrolytic, nutritional or infectious aetiologies of cardiac dysfunction were eliminated. The hypothesis of a hypoxic origin then remained.

Two months after his arrival in France, the patient was brought to us urgently by his host family due to a severe hypoxic episode with desaturation at 40%. The intraventricular thrombus was still in place, and we construed this episode as an acute decompensation of its advanced chronic pathology, without identifying any triggers. After stabilization in the intensive care unit, a palliative rescue intervention was decided upon. We performed a systemic-to-pulmonary shunt by sternotomy, in the form of a modified Blalock–Taussig shunt (BT shunt) with an undersized Gore-Tex tube (5 mm in diameter). Anticoagulants, antiplatelet agents and pulmonary vasodilator treatment were initiated.

Echocardiography 12 days after surgery showed significant improvement in biventricular function (Video 2).

Eighteen days after surgery, the child’s condition deteriorated again. He was taken to the hospital with 30% blood oxygen saturation and a lactate rate at 15 mmol/l. Echocardiography revealed thrombosis of the BT shunt. Complete surgical valve-sparing repair was carried out urgently.

The postoperative course was free from complications. Ventricular function improved to a left ventricular ejection fraction of 55%. After 1 month of postoperative supervision, the patient was able to return to his country of origin.

DISCUSSION

The literature concerning these rare forms of TOF with severe biventricular dysfunction is poor. We found 1 case report from 1997 describing a similar case [3]. However, the different surgical
teams contacted during patient management agreed that prognosis was bleak. Several of them advised us to ‘return this patient to his parents without surgery but alive’.

Initial blood oxygen saturation was relatively good for a late presentation of TOF, prompting us to look for other aetiologies of cardiac dysfunction. Improvement in ventricular function after BT shunt suggested a hypoxic origin.

A systemic-to-pulmonary shunt increases the blood flow to pulmonary circulation and the left ventricle preload. In a patient with impaired left ventricular function, this can be risky, but we were forced to do so by the deterioration of the patient’s clinical condition. It should be noted that undersized shunt was well-tolerated and allowed recovery of biventricular function. Pulmonary vasodilator treatment probably allowed the vascular bed downstream of the shunt to be opened, avoiding pulmonary oedema. A normal-sized shunt would have further overloaded both the pulmonary circulation and left ventricle, which would certainly have been deleterious.

Thrombosis of BT shunt resulted in complete repair. It was initially planned a few months from the first surgery for optimal preoperative recovery.

CONCLUSION

Chronic cardiac hypoxia may be responsible for severe biventricular dysfunction in late presentations of TOF. An undersized BT shunt is well-tolerated and allows for a fast recovery of the ventricular function. This recovery enables a delayed but complete surgical repair.

Conflict of interest: none declared.

REFERENCES