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Brief Report

Extracorporeal membrane oxygenation as a bridge to surgical treatment of flail tricuspid valve in a neonate

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Abstract A term infant rapidly developed profound cyanosis and metabolic acidosis shortly after an uncomplicated vaginal delivery. Echocardiography identified a flail antero-superior leaflet of the tricuspid valve, which was producing severe tricuspid insufficiency. The clinical state deteriorated despite maximal medical management, and the patient was placed on venoarterial extracorporeal membrane oxygenation. Within twenty-four hours, the metabolic acidosis corrected, inotropic support was discontinued, and the patient was weaned to minimal ventilator settings. Successful repair of the tricuspid valve was performed two days later.

Keywords: Atrioventricular valve; anterior papillary muscle; extracorporeal membrane oxygenation; congenital heart disease

Myocardial dysfunction resulting in tricuspid regurgitation is commonly associated with neonatal asphyxia.[1–3] Although this may be fatal, the usual findings are transient dysfunction of the myocardium and cardiac valves.[2] In autopsy studies of newborns without congenital heart disease, approximately one-third had evidence of subendomyocardial necrosis, often localized to the region of the anterior papillary muscle of the tricuspid valve.[4,5] We are aware of at least two case reports that describe infants dying from cardiac failure caused by rupture of a papillary muscle and flail tricuspid valve.[6,7] Herein, we describe the striking case of an infant with no apparent antenatal or perinatal distress who developed severe cardiac failure on the first day of life due to flail antero-superior leaflet of the tricuspid valve. The infant was stabilized using extracorporeal membrane oxygenation, and underwent successful repair of the tricuspid valve.

Case report

An infant of 38 weeks gestational age, who had undergone a normal prenatal ultrasonic interrogation at 29 weeks of gestation, was delivered vaginally with APGAR scores of 8 at 1 and 5 minutes, 2 points having been deducted because of abnormal colour. On physical examination, there was generalized cyanosis, a holosystolic murmur, and hepatomegaly. Saturations of oxygen ranged from 20 to 50%, and the chest x-ray demonstrated normal cardiac size with decreased pulmonary vascularity.

Echocardiography (Fig. 1a,b) demonstrated a flail antero-superior leaflet of the tricuspid valve, with thickening at the tip of the leaflet, an unusual echodensity at the apex of the right ventricle, severe tricuspid insufficiency indicating right ventricular pressures to be at systemic levels, right atrial enlargement with right-to-left shunting through a patent oval foramen, a small patent arterial duct permitting left-to-right shunting, and normal biventricular function.

The patient developed persistent hypotension and metabolic acidosis despite the institution of high-frequency oscillator ventilation, nitric oxide, fluids in excess of 250 millilitres per hour, dopamine at 25 micrograms per kilogram per minute, dobutamine at 20 micrograms per kilogram per minute, and a stress dose of hydrocortisone. Given the lack of clinical response, we initiated venoarterial extracorporeal membrane oxygenation.

Within 24 hours, the metabolic acidosis corrected, inotropic support was discontinued, and the patient

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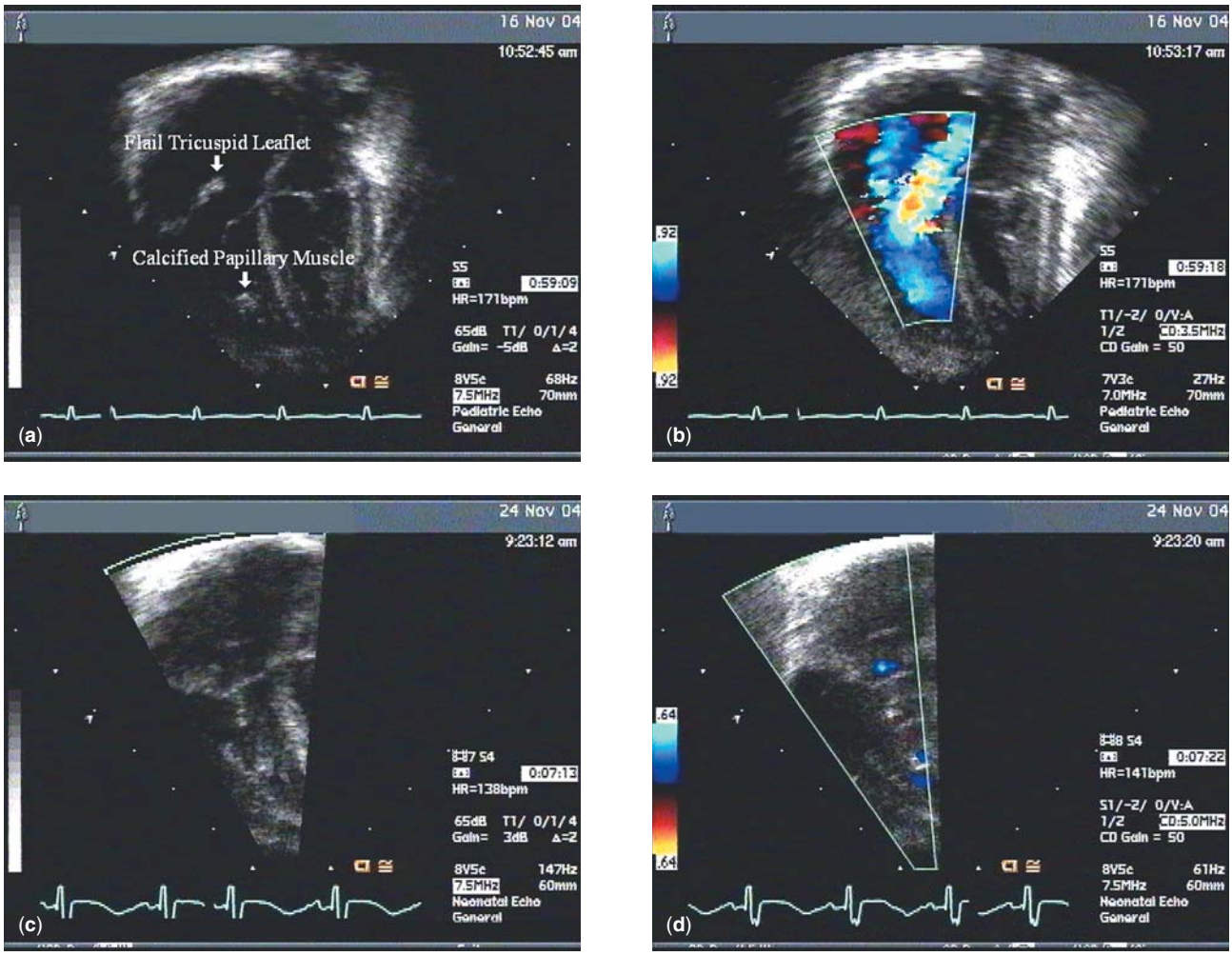

Figure 1.
*Pre-operative echocardiogram demonstrating (**a**) flail antero-superior leaflet of the tricuspid valve with thickening at the tip of the leaflet, calcified papillary muscle at the apex of the right ventricle, right atrial enlargement with bowing of the interatrial septum to the left and (**b**) severe tricuspid regurgitation. The post-operative echocardiogram demonstrates (**c**) excellent systolic coaptation of the re-suspended antero-superior leaflet with the septal leaflet and (**d**) only trivial insufficiency.*

was weaned to minimal ventilator settings. The patient was decannulated after 5 days of extracorporeal membrane oxygenation, and placed on a conventional ventilator.

At surgery, two days later, inspection of the tricuspid valve demonstrated that the anterior papillary muscle had ruptured, causing the flail antero-superior leaflet. New tendinous cords were created by resuspending the leaflet to the base of the anterior papillary muscle and the annulus was down-sized with a Kaye annuloplasty. Transesophageal echocardiography demonstrated excellent systolic coaptation of the leaflets, no stenosis, and trivial insufficiency. Repeat echocardiography on the seventh post-operative day, and at three months of life, demonstrated no tricuspid stenosis with trivial insufficiency (Fig. 1c,d).

Discussion

One of the most common causes of cardiac failure on the first day of life in neonates with a structurally normal heart is myocardial dysfunction due to asphyxia during birth. The anterior papillary muscle of the tricuspid valve is particularly vulnerable to ischaemic injury[1–3] because of its high requirement for oxygen, perfusion being possible only during diastole in the setting of systemic right ventricular pressures, and its location at the distal extreme of the coronary circulation.[4] As a consequence, perinatal hypoxia can cause congenital tricuspid regurgitation, and even flail tricuspid valve. This was the situation in our patient, who suffered severe congestive cardiac failure due to rupture of the anterior papillary muscle. In our case, the aetiology for the rupture is unknown, but most

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likely resulted from an unrecognized period of intrauterine asphyxia shortly before birth. Other less likely aetiologies include traumatic rupture during the birth process, or a thromboembolic event.

In contrast to most patients with Ebstein's malformation, flail tricuspid valve results in severe cardiac failure within the first few hours of life due to the sudden onset of severe tricuspid insufficiency. As manifest in this, and other reported cases, there is rapid cardiovascular decompensation refractory to conventional anti-congestive therapy, and a high risk of death.[6,7] The difference is probably due to the abrupt decrease in stroke volume associated with flail tricuspid valve, compared with relatively compensated cardiac failure encountered in the setting of Ebstein's malformation due to slow cardiac enlargement during fetal life. Extracorporeal membrane oxygenation has proven a reliable mode of cardiorespiratory support in neonates and children with presumptively lethal cardiac dysfunction,[8–10] and was lifesaving in our patient. Neonatal flail tricuspid valve results in severe and rapidly progressive haemodynamic instability, and extracorporeal membrane oxygenation was crucial to survival. Given the lack of response to conventional therapy, we suggest that extracorporeal membrane oxygenation should be considered early in the management of infants with flail tricuspid valve.

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