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Brief Report

Successful repair of critical tricuspid regurgitation secondary to ruptured papillary muscle in a newborn

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Abstract We report a rare case of an isolated critical tricuspid regurgitation due to rupture of a papillary muscle. This patient presented with a cyanosis immediately after birth. Despite mechanical ventilation and medical management to decrease the pulmonary vascular resistance, the low cardiac output persisted, along with the cyanosis. Repair of the tricuspid valve was performed, using an artificial tendinous cord, on the 4th day of life. The short-term result of the surgery is satisfactory, but the patient requires long-term follow-up.

Keywords: Atrioventricular regurgitation; artificial cords; cyanosis

Isolated critical tricuspid regurgitation in neonates is a rare and potentially lethal disease. The causes of regurgitation vary from dysplasia of the leaflets to rupture of the tension apparatus. Although the incidence of rupture of a papillary muscle is very low, it should be considered as a possible cause when hypoxemia is encountered in a newborn along with severe tricuspid regurgitation. The outcome of such massive tricuspid regurgitation in a newborn is generally poor, even in cases with surgical intervention. We diagnosed a 2-day-old neonate with isolated critical tricuspid regurgitation caused by rupture of a papillary muscle. He underwent successful surgical repair using an artificial tendinous cord inserted during the early neonatal period.

Case report

A 2-day-old boy was transferred to our hospital due to severe cyanosis and tachypnea. He was born at term by a Cesarean section, with a weight of 3000 g. He had been doing well until 10 days prior to birth, when his mother noticed a decrease in fetal movements. Soon after birth, the baby developed severe cyanosis. Although ventilatory support was provided, he remained profoundly hypoxemic and he was then transferred to our hospital. On admission, he had cyanosis, dyspnea and tachypnea. The precordium was hyperactive, and a harsh pansystolic murmur of grade 3/6 was heard at the lower left sternal border. The hepatic margin was palpated three fingers below the right costal margin. The peripheral pulse was somewhat weak. Chest radiography revealed oligemic lung fields and there was marked cardiomegaly. An echocardiogram showed severe tricuspid regurgitation, with a flail antero-superior leaflet with a thickened, echogenic tip (Fig. 1).

The right atrium was markedly dilated. A right-to-left shunt was detected through the patent oval foramen. Systolic function of both ventricles was favorable.

Despite maximum conventional ventilatory support, and subsequent high frequency oscillatory ventilation, the infant continued to be severely hypoxemic. Other adjunctive measures, including infusion of sodium bicarbonate, inotropic agents, diuretics, muscle relaxant and prostaglandin E1, all failed to improve either oxygenation or the state of low cardiac output. Continuous inhalation of nitric oxide, aimed at lowering the pulmonary vascular resistance was tried and this produced a minimal increase of oxygenation. On the 4th postnatal day, we weaned him from nitric oxide because of his

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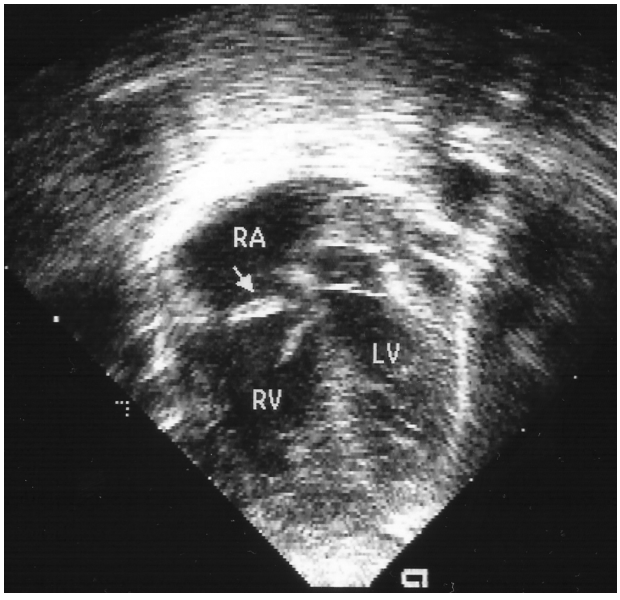

Figure 1.
The preoperative transthoracic echocardiogram shows a flail anterosuperior leaflet of the tricuspid valve with a thickened, echogenic, tip prolapsing into the right atrium (arrow). RA: right atrium; RV: right ventricle; LV: left ventricle.

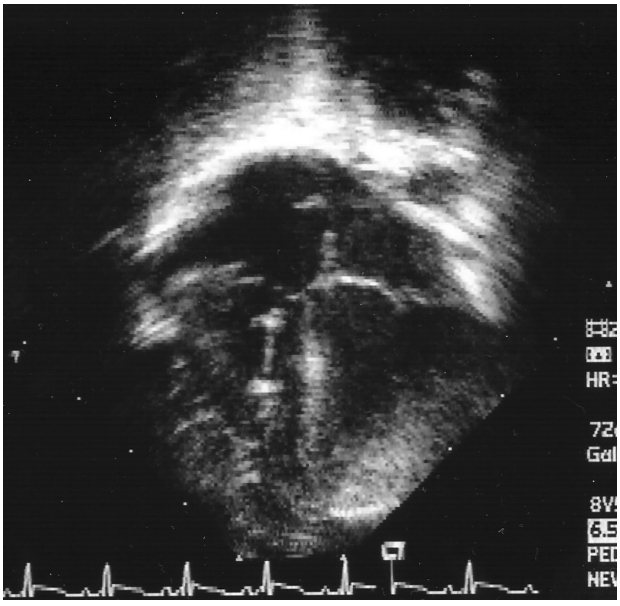

Figure 2.
The postoperative transthoracic echocardiogram shows the artificial cord supporting the leaflet and restoring adequate coaption.

methemoglobinemia. After the discontinuation of nitric oxide, his cyanosis became profound and his serum lactate level increased to 4.4 mmol/l. The serious condition of the patient led us to attempt a surgical repair of the tricuspid regurgitation. We discovered that the anterior papillary muscle was ruptured and calcified, with additional mild calcification of the inferior papillary muscle. During the operation, we excised the anterior papillary muscle, and created an artificial cord from plegeted 7-0 Gore-Tex. In addition, we carried out a tricuspid annuloplasty.

After the repair, his cyanosis disappeared, and cardiac output improved dramatically. A postoperative echocardiogram showed minimal to mild tricuspid regurgitation, a normally sized right atrium, and good systolic function of both ventricles (Fig. 2).

He was discharged on the 15th postoperative day. After eight months of postoperative follow-up, he remained very well, and no cardiac murmur was audible. At that time, his echocardiogram showed normal cardiac function with minimal tricuspid regurgitation.

Discussion

Congenital tricuspid insufficiency is uncommon, but is a recognized cause of neonatal congestive heart failure, producing massive cardiomegaly and cyanosis. These clinical findings have been found even in neonates who do not have primary tricuspid abnormalities. In general, it is known to be associated with prenatal or perinatal myocardial damage in the stressed newborn.[1] Concerning the location of myocardial hypoxic damage, Donnelly et al.[2] demonstrated that stressed neonates are at high risk of having ischemic myocardial necrosis, especially in the papillary muscles. Tricuspid valvar insufficiency secondary to the rupture of the papillary muscle has previously been reported by Alkalay et al.[3] Their patient presented with fetal distress. At the age of 12 h, the newborn died of cardiorespiratory failure. It was suggested that the prenatal hypoxic insult was related to rupture of the papillary muscle. In addition to that case, two other neonatal cases were reported respectively.[4,5] Both neonates had perinatal hypoxia, and they needed resuscitation. Neither responded to medical management and both infants died.

Medical treatment, which is aimed at reducing the high pulmonary vascular resistance, allows for an increase of antegrade flow from the right ventricle and also should improve the systemic perfusion.[6] Should the clinical state of the neonate continue to deteriorate, however, and no further improvement is established, any plastic procedure must be considered to minimize the backflow through the tricuspid valve. Repair using artificial tendinous cords has been reported in children by Reddy et al.[7] Cordal replacement, or augmentation with an expanded polytetrafluoroethylene suture, was shown to be a useful technique when the congenitally dysplastic valves had abnormal cordal support. To the best of our knowledge, however, no repair of this type has been attempted in a neonate.

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In our case, the etiology of the rupture is unclear. Although no obvious evidence for perinatal hypoxia was obtained, a decrease in fetal movement before the delivery might be related to myocardial damage. In addition, the flail valve was initially interpreted to be the result of a ruptured tendinous cord from the anterior muscle, and urgent surgical intervention was deemed necessary. Surprisingly, the short-term result of the surgery was excellent. The success of the treatment was mainly attributed to the early surgical management for the correction of valvar regurgitation. Until recent times, surgery on these neonates is known to carry a high mortality.[8–10] Because of this, surgery is not usually recommended. In considering the recently improved surgical outcomes for neonatal surgery, it should be emphasized that early surgical intervention may increase the chances of survival when medical management is proving ineffective. Although there may be a need for further operations as our child grows, it could be also possible that compensatory growth of the leaflet will be sufficient to maintain an adequate valvar function. On the basis of our limited experience, we conclude that artificial replacement of tendinous cords could become an acceptable approach to repair of the tricuspid valve in newborns with regurgitation due to ruptured papillary muscles.

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