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Brief Report

Right ventricular noncompaction in a neonate with complex congenital heart disease

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Abstract Ventricular noncompaction is a rare unclassified cardiomyopathy occurring because of arrest of the normal intrauterine compaction of the loose luminal component of the ventricular myocardium. There is limited data regarding its diagnosis and outcome in children. It is recognised, however, that right ventricular involvement is extremely rare. We report a case in which only the right ventricular myocardium was noncompacted, a situation which led to heart failure soon after birth.

Keywords: Cardiomyopathy; echocardiography; cardiac failure

During the early phase of embryogenesis, the ventricular myocardium possesses compact and trabecular layers. As development proceeds, the trabecular layer becomes compacted, and loses its initial mesh-like pattern. Failure of compaction is now recognized as a cardiomyopathy, albeit as yet unclassified. It is characterized by prominent trabeculations and deep intertrabecular recesses, especially in the left ventricle, which at times also affects the right ventricle and the muscular ventricular septum. Right ventricular involvement in the neonate, however, is extremely rare. We report a 1-day-old infant in whom right ventricular noncompaction was recognized using echocardiography.

Case report

The propositus was born at term after an uncomplicated gestation, being delivered in another institute. The weight at birth was 3,300 grams, representing the 50th centile, length was 51 centimetres, at the 75th centile, and head circumference was 35 centimetres, again at the 75th centile. Soon after birth, the neonate developed cyanosis, with an initial capillary partial pressure for oxygen of 28 torr, nH of 7.15, and saturation of oxygen of 60 per cent in room air. The infant was referred to our institution because her clinical state was not improved despite the institution of medical treatment. At the initial examination, the infant was noted to be severely cyanotic and agitated, with a heart rate of 120 beats per minute. Respiratory rate was 64 breaths per minute. A systolic murmur, graded at 2 out of 6, was heard at the left sternal border. Chest radiography revealed the heart to be enlarged, but there was no pulmonary congestion. Laboratory data included an hematocrit at 53 per cent, red blood cell count of 4.97×10^6 per millilitre, white blood cell count of 15×10^3 per millilitre with 42 per cent segmented forms, and 55 per cent lymphocytes. The neurologic state was normal, with good muscle tone.

Echocardiographic interrogation revealed an atrioventricular septal defect with common atrioventricular junction, the common valve being committed almost exclusively to the right ventricle, with severe hypoplasia of the left ventricle. There was a common atrium (Fig. 1). The aorta arose anteriorly from the right ventricle, and there was pulmonary atresia, with hypoplastic intrapericardial pulmonary arteries perfused via systemic-to-pulmonary collateral arteries. The interrogation also revealed huge trabeculations and deep recesses in the ventricular myocardium, which were confined to the right ventricle, which functionally was the systemic ventricle (Figs 1 and 2). On the basis of these findings, we diagnosed

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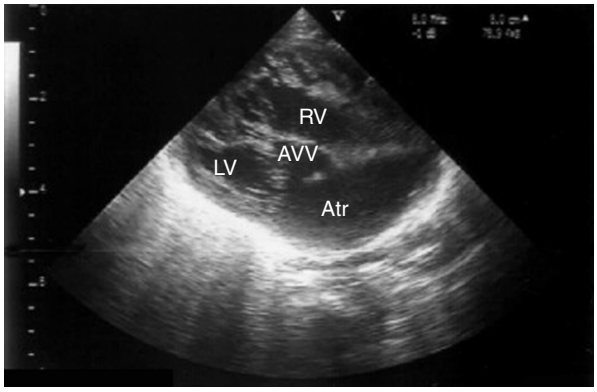

Figure 1.
Cross-sectional echocardiograms in the infant with right ventricular noncompaction. The parasternal long-axis view clearly demonstrates a thin compact layer of the right ventricular wall, and an extremely thickened trabecular layer with prominent trabeculations and deep recesses. Note the common atrioventricular valve (AVV) draining the common atrium (Atr) predominantly to the right ventricle (RV), the left ventricle (LV) being hypoplastic.

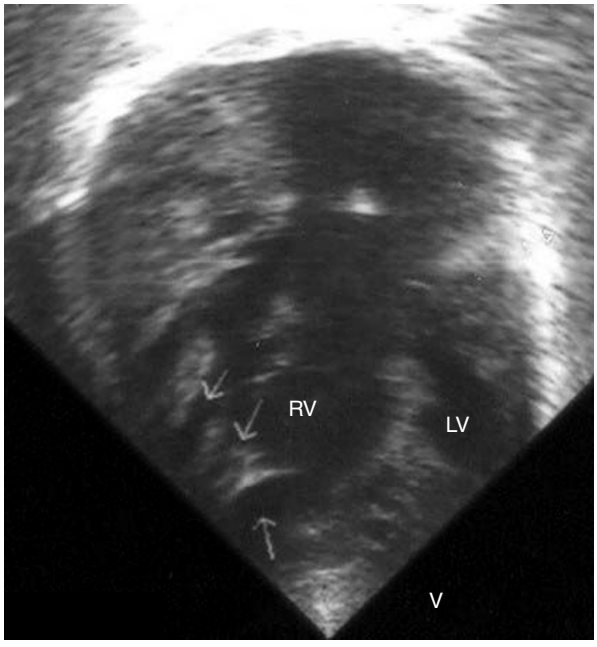

Figure 2.
The parasternal four-chamber view confirms the presence of prominent trabeculations and deep recesses (arrowed) in the right ventricle (RV). The section also shows the hypoplastic left ventricle (LV) and the common atrium.

noncompaction of the right ventricle in association with complex congenital cardiac disease. Right ventricular function was depressed, with a fractional shortening of 17 per cent. Cardiac catheterization confirmed the diagnosis. Pressures in the right ventricle were measured at 70 millimetres of mercury systolic, at 2 millimetres of mercury as a mean in the right atrium, with systolic pressure of 68 in the aorta, the diastolic pressure being 37 millimetres of mercury, and the mean pressure 51 millimetres of mercury. The infant was treated with digitalis and diuretics. She was discharged from hospital at the age of 7 days with a good clinical condition. Unfortunately, the infant died suddenly at home at the age of 15 days. We were unable to obtain a postmortem examination.

Discussion

Isolated noncompaction of the left ventricle is characterized by the presence of numerous prominent trabeculations and deep intertrabecular recesses within the luminal layer of the ventricular myocardium.[1–3] The abnormality is reported to be accompanied by three major risks, namely depressed ventricular function, systemic embolization, and ventricular arrhythmias. Due to the depressed ventricular function, patients with the abnormality usually present with heart failure similar to that of dilated cardiomyopathy. Right ventricular involvement is extremely rare,[4,5] with only limited reports of right ventricular noncompaction being recognized in neonates.[6]

In the early period of embryogenesis, the developing ventricular wall is made up of a loose network of myocardial fibres that normally condenses to produce compact and trabecular layers, with disappearance of large intertrabecular spaces characterizing the initial phases of development.[3] Noncompaction is believed to reflect an arrest in the normal process of compaction. It may occur in isolation, or may be associated with other congenital cardiac malformations.[7] It is usually the isolated forms of left ventricular noncompaction which thus far have been reported in the literature.[1–3,7]

When found, the noncompaction involves the majority of the segments of the left ventricular myocardium, making diagnosis relatively straightforward when noncompaction is severe. In the normal heart, however, the apical component of the right ventricular possesses prominent coarse trabeculations. This normal echocardiographic feature of the right ventricle makes it difficult to differentiate the normal from pathologic patterns of noncompaction. Prominent trabeculations, coupled with hypokinesia of the right ventricle, in the setting of obvious left ventricular noncompaction, may permit the diagnosis of right ventricular involvement. In a large series of patients reported with noncompaction of the left ventricular myocardium, right ventricular involvement was reported in two-fifths of cases, with left ventricular noncompaction also being seen in association with other congenital cardiac malformations. In our patient, however, the left ventricle was

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extremely hypoplastic, giving a functionally single right ventricle. Due to this, we were unable to identify the pattern of trabeculation in the left ventricle. So-called "myocardial sinusoids" are known to be associated with congenital obstructive lesions, but always in the setting of an intact ventricular septum. Although our patient had atresia of the pulmonary trunk, there was no obstruction to right ventricular outflow tract, since the unobstructed aorta was arising exclusively from the right ventricle. We do not think, therefore, that the pattern of noncompaction seen in our patient was due to pulmonary atresia in itself. At all events, the findings of so-called "sinusoids" seen in pulmonary atresia are fundamentally different from the pattern increasingly being recognized as representative of right ventricular noncompaction.

Noncompaction of the ventricular myocardium, therefore, is a rare congenital cardiomyopathy that may manifest itself in the early neonatal period. Echocardiographers need to be familiar with the diagnostic pattern of noncompaction in order to prevent any delay in diagnosis. Our findings of noncompaction in the early neonatal period support the concept that the lesion is caused by an arrest of normal myocardial compaction during the fetal period, and that it can cause severe right heart failure subsequent to birth.

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