Noncompaction of the right ventricle following Senning repair

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A three-year-old boy presented with generalized edema, respiratory distress, prominent right ventricular impulse and hepatosplenomegaly. He had undergone Senning repair at one year of age. On his echocardiography, there were numerous prominent trabeculations and deep intertrabecular recesses measuring approximately 5 mm in depth along the free wall and right ventricular apex. Echogenicity of the endocardial surface was increased suggesting a fibrotic process in progress. Intertrabecular recesses were observed to be filling from the ventricle by color Doppler which is consistent with noncompaction. Various semilunar valve obstructions were shown to be responsible for the persistence of deep endomyocardial spaces surrounded by exeggerated hypertrophy of the trabeculae. This report presents the echocardiographic findings of right ventricular cardiomyopathy associated with dextroposition of the great arteries following Senning operation resembling noncompaction. Thus, this rare entity needs to be clarified regarding morphological criteria in distinction from other cardiomyopathies.

Key words: Senning repair, cardiomyopathy, noncompaction.

Postnatal persistence of spongy myocardium with embryonic blood supply may accompany a number olf congenital defects. Deep intertrabecular recesses and prominent trabeculae are features of the embryonic or spongy myocardium that normally develops into the compact myocardium characteristic of the mature mammalian heart[1-11]. Noncompaction has been reported to occur as a form of hypertrophic or restrictive cardiomyopathy and endocardial fibroelastosis[12,13]. We describe the typical echocardiographic appearance of isolated noncompaction in a postoperative case with the diagnosis of dextroposition of the great arteries.

Case Report

A three-year-old boy presented with swelling of the face and extremities and respiratory distress. He had undergone Senning procedure at one year of age with the diagnosis of dextroposition of the great arteries. Permanent pacing had been initiated approximately six months after surgery due to persistent dysrhythmias. Following hospital admission, hyperbilirubinemia developed due to hepatitis B virus. When he first presented he was already on digoxin and diuretics. He was tachypneic and in respiratory distress. There was generalized edema with hepatosplenomegaly. Right ventricular impulse was prominent. There was cardiomegaly and right ventricular hypertrophy on his chest X-ray and electrocardiogram (ECG). An echocardiogram was performed with HP-1000 Sonos and 3.5 mHz transducers. There was no obstruction to the pulmonary venous flow. Both ventircles were enlarged. Systolic function was depressed. There were numerous prominent trabeculations and deep intertrabecular recesses measuring approximately 5-7 mm in depth along the free wall and apex of his right ventricle (RV). RV free wall was thickened (Fig. 1). Echogenicity of the endocardial surface was increased, suggesting a fibrotic process in progress. Intertrabecular recesses were observed to be filling from the ventricle by color Doppler, which is consistent with noncompaction. Pulmonary and 1 (+) tricuspid regurgitation were present. The patient died at home shortly after his discharge.

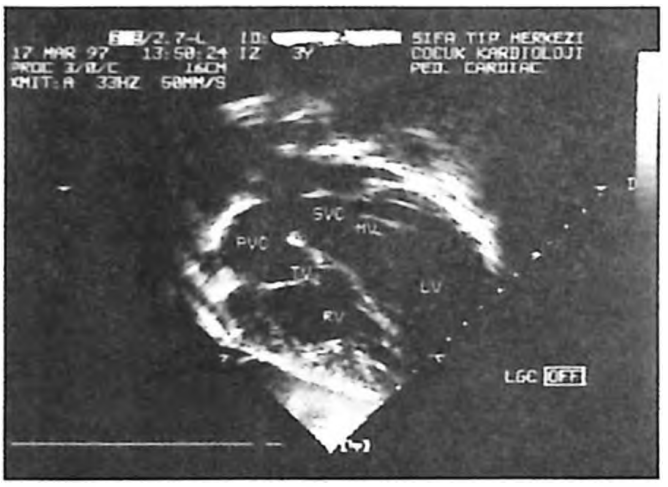

Fig. 1. Two-dimensional echocardiographic appearance of right ventricular cardiomyopathy.

Discussion

Right ventricular dysfunction may occur in the long-term follow-up of patients undergoing atrial switch[1-5]. In a prospective study, the majority of patients,with transposition of the great arteries who underwent Mustard repair were in NYHA classes I and II at a mean follow-up period of 18 years, with mean RV ejection fraction values of 39%. Wilson et al.[6] reported that only half of the patients were in sinus rhythm; one patient was in complete heart block and the rest were in junctional rhythm. High occurrence of early and late postoperative dysrhythmias is more likely to cause injury to the sinoatrial node and/or its arterial supply than disruption of internodal tracts or damage to the atrioventricular node[6]. This case presented manifested persistent dysrhythmias requiring permanent pacemaker implantation. Neither shunting across the intraatrial repair nor obstruction to systemic or pulmonary venous return was present. However, RV and left ventricular (LV) dysfunction must have led to marked dilatation of both ventricles. It seems likely that one of the problems was the RV fuctioning as the systemic pumping chamber.

Various forms of bizarre trabecular patterns may be noted on angiocardiograms in patients with complex cyanotic congenital heart disease[7-9]. Failure of the normal differentiation of the primitive ventricular wall into the compact myocardium has been proposed in the mechanism of the abnormal patterns seen in some congenital heart diseases. With the advence of noninvasive techniques, a new entity, isolated noncompaction, has been described recently by echocardiography and confirmed pathologically. Diagnostic criteria for isolated noncompaction are prominent trabeculations associated with deep intertrabecular recesses. However, it must be noted that discrete muscle bundles, three or less in number and more than 2 mm in diameter, have been reported in 68% of normal hearts[10]. It was proposed that in isolated noncompaction increased fibrous and elastic tissue on the endocardium and within the intertrabecular recesses may be due to subendocardial ischemia[10]. The arrhythmogenic aspect observed in LV noncompaction has been explained by the morphological appearance of the troughs of the intertrabecular recesses being reminiscent of RV dysplasia. However, in a larger series reported, isolated noncompaction was not considered comparable to arhythmogenic RV dysplasia because the morphologic characteristics of these conditions differ completely[11]. Both studies reported that the intertrabecular recesses showed continuity with the left ventricular cavity. This finding has been depicted as an expression of an arrest of compaction of the ventricular myocardium, more appropriately terming the disorder noncompaction, rather than persisting sinusoids. Abnormal mitochondrial structure and mitochondrial chain abnormalities have been established in noncompaction associated with ventricular septal defects. Isolated noncompaction was reported to present as restrictive cardiomyopathy and endocardial fibroelastosis[12]. The cardiomyopathy observed in this case was distinct from arrhythmogenic RV dysplasia, which is characterized anatomically by patchy or diffuse adipose or fibroadipose substitution of RV myocardium and akinetic areas with discrete wall bulges in the absence of any other identifiable structural heart or pulmonary disease[13]. Neither does it resemble Uhl's anomaly, which is marked by extreme thinning of the RV, which may rarely be seen as an associated pathology in patients with congenital left or right ventricular obstruction and with anomalous origin of the left coronary artery from the pulmonary trunk. Although histopathological examination was not possible in this case, the echocardiographic findings were identical to the features of isolated noncompaction described by Chin et al.[10] Isolated noncompaction has been previously decribed as involving either both ventricles or the LV alone, but never just the RV[11].

To our knowledge, an echocardiographic appearance resembling that of noncompaction as a form of cardiomyopathy of the right ventricle following Senning operation has not been reported to date. This rare entity needs to be clarified regarding morphological criteria to distinguish it from other cardiomyopathies.

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