Biventricular noncompaction in a patient with dextrocardia/dextroversion diagnosed with cardiac magnetic resonance imaging

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A five-year-old boy was referred for follow-up of a surgical correction of partial anomalous pulmonary venous return. Echocardiography and magnetic resonance imaging (cMRI) showed visceral heterotaxy with bilateral left atrial appendage morphology. The ventriculoarterial connection was concordant with dextrocardia/dextroversion. Multiple muscular ventricular septal defects near the apex were also observed. There were no ventricular outflow tract or aortic arch obstructions. A slightly increased flow was observed in the repaired pulmonary veins but there was no evidence of obstruction. The patient's left ventricle (LV) demonstrated extensive trabeculations with flow into the deep intertrabecular recesses, which suggested the presence of LV noncompaction (Figure 1). cMRI confirmed the diagnosis of myocardial noncompaction in the LV (Figure 2). Most of the LV was affected, except for the basal part of the septum, with a noncompacted to compacted layer ratio of 4:1. The involved area showed significant

Figure 1) Colour Doppler echocardiogram in parasternal short-axis view, showing penetration of colour into the deep intertrabecular recesses

thinning of the compact layer of the myocardium. Furthermore, there was involvement in the apical part and inferior wall of the right ventricle (RV) (Figure 2).

Despite these findings, systolic function was within normal limits and the patient was asymptomatic. The patient continues to follow up on a regular basis. He remains off medications and has a normal level of activity with no symptoms or signs of heart failure, arrhythmia or thromboembolic events.

Figure 2) Short-axis fast cine magnetic resonance image of the ventricles shows typical lacy pattern of noncompacted left ventricular (LV) myocardium (arrows). Note that the LV free wall is more extensively involved than the ventricular septum. The compact layer is markedly thinned in the involved area. The right ventricle (RV) also shows a similar trabeculation pattern

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Noncompaction of the ventricles is a newly recognized cardiomyopathy (1) that is believed to arise from arrested endomyocardial development during embryogenesis. The incidence of this cardiomyopathy in all pediatric patients receiving echocardiograms is 0.05%, but was found to be 2% in those patients with congenital heart disease (up to 22% in patients with single left ventricle) (2).

Noncompaction has primarily been described in the LV; however, there have been reports of biventricular or isolated RV involvement (3). Because of increased trabeculations normally found in the RV, RV noncompaction is more difficult to diagnose than LV noncompaction.

Ventricular noncompaction has been most extensively studied using two-dimensional and colour Doppler echocardiography. cMRI is increasingly being used to more accurately diagnose LV noncompaction. Borreguero et al (4) suggested the potential role of cMRI in the evaluation of the RV with noncompacted myocardium. Our patient represents an example of biventricular noncompaction in which cMRI aided the diagnosis. To our knowledge, the present patient is the first to be described with biventricular noncompaction in the presence of dextrocardia/dextroversion since the official description of this cardiomyopathy. A case with a similar biventricular "bizarre trabecular pattern" was published almost 30 years previously (5). However, in that case, there was no clear description regarding the extension and the ratio between the compacted and

noncompacted myocardium. Furthermore, that case was associated with anomalies that generate significant intraventricular pressure overload.

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