Isolated Left Ventricular Apical Hypoplasia

We present the case of a 9-year-old asymptomatic female, in whom a systolic murmur in the mesocardia was detected upon routine examination. The echocardiogram showed abnormal morphology of the right ventricle (RV). Magnetic resonance (MR) showed the RV wrapped around the left ventricle (LV) at the cardiac apex (Figures 1 and 2). Figure 1 is a 4-chamber view in diastole that shows the reduction in the longitudinal diameter of the LV due to the lack of development of its apical portion and the substitution of fatty tissue (arrow), characteristic of dysplasia affecting this area. Figure 2 is a coronal oblique view that shows the RV shaping the cardiac apex and (arrow) the accentuated ventricular concavity over the LV. Figure 3 is a 4-chamber view in systole; the arrow indicates the lack of contraction by the fatty tissue, which protrudes toward the apex of the right ventricular cavity as the left intraventricular pressure increases.

Differential diagnosis of this pathology requires the presence of left ventricular hypoplasia, aneurysms and ventricular diverticula. The ventricular hypoplasia is characterized by a small and hypertrophic ventricular chamber and dysplastic cardiac valves. The diverticula originates on the ventricular wall at a narrow joint and are more frequent in the LV. Ventricular aneurysms are acquired defects, located at a wide junction, that buckle during systole.

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