**ISOLATED FAMILIAL RIGHT VENTRICULAR HYPOPLASIA**

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Right ventricular (RV) hypoplasia may develop secondary to pulmonary or tricuspid valve atresia. Those patients are usually symptomatic and need prompt intervention. Isolated RV hypoplasia is a rarely reported congenital heart disease. We report a rare case of 23 year old twins who have been monitored for the last 12 years for an isolated right ventricular hypoplasia. While the boy has been completely non-symptomatic, the girl recently developed some exercise intolerance. Echo and MRI studies (figures) showed a small heavily trabeculated non apex forming RV and a mild tricuspid valve insufficiency. In addition, the girl has a small secundum ASD. Otherwise, the cardiac anatomy and function was normal. The family history was remarkable for their father dying at the age of 30 years and his autopsy was positive for a hypoplastic RV. The paternal uncle also died at age 46 years and his son has an unidentified congenital heart disease. Chromosome microarray analysis of the twins did not identify any copy number variations that are of clinical significance based on the current reporting criteria. The family history appears to suggest an autosomal dominant pattern of inheritance with variable expressivity.

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