Effect of Ser96Ala variant in histidine-rich calcium-binding protein on DCM


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Ebstein anomaly and hypertrophic cardiomyopathy

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An asymptomatic 72-year-old man without cardiovascular risk factors was evaluated due to a systolic murmur on an examination. Transthoracic echocardiogram revealed severe left ventricular hypertrophy associated with Ebstein anomaly (Panel A). An asymmetric septal hypertrophy was observed (thickness = 22 mm). Left ventricular cavity was reduced (Panel B). A peak gradient of 40 mmHg was obtained in the left ventricular outflow tract (Panel C). The tricuspid valve insertion was very distal, close to the apex, with almost complete atrialization of the right ventricle with the exception of a small infundibular component. However, tricuspid regurgitation was no more than moderate. Calculated pulmonary artery systolic pressure was 40 mmHg. Right chambers were not enlarged. A cardiac magnetic resonance was performed confirming the findings (Panels D and E). It showed apical displacement of the hinge point of the septal and posterior leaflet from the atrioventricular ring, Ebstein anomaly was type B, with a large atrialized component of the right ventricle, but the anterior leaflet moves freely (Panel F). The patient had no previous history of supraventricular tachycardias, and familial screening was negative.

Ebstein anomaly is a rare condition (1–5 per 200 000 live births and <1% of all congenital heart defects). It often associates with left heart abnormalities involving the myocardium or valves. The most common finding is a myocardial anomaly resembling mild non-compaction, but the uncommon is the presence of hypertrophic cardiomyopathy.

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