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Circulating Biomarker for Myocardial Angiogenesis in Children and Adolescents with Severe Hypertrophic Cardiomyopathy

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Introduction: Hypertrophic cardiomyopathy (HCM) is characterized by gradual thickening of the myocardium with disruption of coordinated cardiac hypertrophy and angiogenesis resulting in impaired coronary collateralization. Endostatin, an important angiogenesis inhibitor, is a 20-kDa carboxy-terminal proteolytic fragment from the first non-collagenous domain of collagen XVIII, which is a central component of the extracellular matrix in the cardiovascular system.

Objective: To assess in young patients with familial HCM whether plasma endostatin relates to the severity of HCM as defined by the presence of intracardiac cardioverter-defibrillator (ICD).

Methods: Children and adolescents with family history and echocardiographic signs of HCM (n=14; median age 15 years, range 7-27 years) as well as healthy controls (n=21) underwent echocardiography with tissue Doppler imaging, cardiac magnetic resonance (CMR) and peripheral venous blood sampling for endostatin analysis. Half of the HCM patients (n=7; “ICD-HCM”) had earlier received ICD either for primary or secondary prevention for sudden cardiac death (SCD).The remaining HCM patients (“HCM”) were age-matched with ICD-HCM patients.

Results: The left ventricular (LV) thickness and mass, and the mitral septal and lateral E/e´ were significantly higher in the ICD-HCM group than in the other 2 groups ($p<0.01$). Plasma endostatin was higher in the ICD-HCM group ($p=0.09$ vs. controls) and correlated with septal E/e´ ($p=0.006$, $r=0.5$) and with LV septal hypertrophy ($p=0.04$, $r=0.4$). In the subgroup of HCM patients who underwent CMR, there was an inverse correlation between endostatin and myocardial perfusion response to adenosine on CMR ($r=-0.45$, $p=0.08$).

Conclusion: In patients with HCM, the degree of septal hypertrophy, diastolic dysfunction and myocardial perfusion response to adenosine correlate with plasma levels of endostatin, which is elevated in the ICD-HCM group. The findings suggest endostatin as additional biomarker for more severe adverse changes in the myocardium of these patients.