P-146

Microvascular Abnormalities in Coronary and Peripheral Circulation in Hypertrophic Cardiomyopathy of the Young

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Introduction: Sudden cardiac death (SCD) remains the most dreaded consequence of HCM, and may be the first presentation of the disease in the young. Initial assessment of young individuals with myocardial hypertrophy or asymptomatic individuals with HCM heredity is thus essential for further diagnostic and the therapeutic decision making.
Objective: To investigate peripheral vasomotor function and myocardial perfusion in young individuals with HCM and in those at risk for developing this disease.
Methods: Based on heredity or genetic predisposition, adolescents and young adults (median 20 years, range 12-30 years) with familial HCM (HCM; n=10, mean IVS 19.6 mm, PW 11.5 mm), and individuals at risk for HCM (HCM-risk; n=14, mean IVS 10.8 mm, PW 9.3 mm), were compared with healthy matched controls (n=12, mean IVS 10.3 mm, PW 9.6 mm). All underwent assessment with echocardiography, cardiovascular magnetic resonance (CMR), and skin Laser Doppler with iontophoresis of acetylcholine (Ach) and sodium nitroprusside (SNP). CMR was performed at rest and during hyperemia with adenosine 140 mcg/kg/min. Myocardial perfusion (MP) was calculated as the ratio of coronary sinus flow and left ventricular mass (LVM) from CMR.
Results: Both echo and CMR demonstrated significantly greater left ventricular thickness and mass in HCM patients ($p< 0.05$) than in controls and HCM-risk. Compared to controls, cutaneous microvascular responses to Ach were enhanced in HCM and HCM-risk ($p<0.05$). MP (ml/min/g) at rest was similar in controls, HCM-risk and HCM patients (0.8±0.1, 1.0±0.1, 0.9±0.1; p=ns). During adenosine-induced hyperemia, HCM patients showed lower MP (2.5±0.4, $p<0.05$) than controls (3.9±0.3) and HCM-risk (5.0±0.5).
Conclusion: These results indicate that microvascular disease may be an early manifestation of HCM in the young.