Exercise Stress Echocardiography in Children with Hypertrophic Cardiomyopathy

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Background: Hypertrophic Cardiomyopathy (HCM) is the most common genetic cardiomyopathy and the leading cause of sudden death in young athletes. The disease commonly causes left ventricular (LV) diastolic dysfunction, and late LV systolic dysfunction. Diastolic dysfunction has been described limiting exercise capacity but less information is available on systolic contractile reserve in pediatric patients. The aim of the current study was to evaluate myocardial response to exercise in children with HCM using semi-supine cycle ergometry stress echocardiography (SSCE).

Materials and Methods: Seventeen children with HCM and 24 controls were included. A stepwise SSCE stress echocardiography protocol was used. Tissue Doppler Imaging (TDI) peak systolic and early diastolic velocities were measured in the LV lateral wall and basal septum during exercise and the changes in E’ and S’ values from baseline to peak were compared between HCM and controls. LV myocardial acceleration during isovolumic contraction was measured in all the subjects at incremental HR to evaluate the force-frequency relationship (FFR).

Results: The change in E’ and S’ from baseline to peak exercise was lower in HCM patients than in controls in LV later wall ($\Delta E'$ 5.7±1.7 vs. 7.5±2.9 cm/s, p=0.03; $\Delta S'$ 3.4 ±4 vs. 7.4 ±3.4 cm/s, p=0.001) and basal septum ($\Delta E'$ 5.1±2.9 vs.6.9 ±1.8cm/s, p=0.02; $\Delta S'$ 3.7±2.3 vs. 5.3±1.6 cm/s, p=0.01). The contractile response as studied by the FFR, was significantly blunted in HCM compared with controls, p=0.02 (see figure).

Conclusions: Our data suggest a significantly blunted myocardial response exercise in patients with HCM. Apart from a previously described reduced diastolic response suggested by the blunted increase in E’-velocities, we also demonstrate a reduced systolic response as suggested by the reduced S’ response and particularly by the blunted force-frequency response.