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 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.

Aims
• The aim of the current study was to evaluate myocardial response to exercise in children with HCM using semi-supine cycle ergometry (SSCE) stress echocardiography

Methods
• Single-center retrospective clinical study approved by the institutional REB
• Two groups of patients:
  - 17 children with HCM
  - 24 age and gender matched controls
• Inclusion criteria: height above 140 cm and normal LV systolic function defined as ejection fraction (EF) greater than 55% at rest.
• Exclusion criteria: physical inability to perform a SSCE test and poor echocardiographic windows at rest
• TDI and deformation analysis was performed using EchoPac (GE Medical Systems, USA)

Results
• At rest:
  - E' values were reduced in HCM group in the LV lateral wall (7.2 ± 3.9 vs. 13.4 ± 2.6 cm/s, p<0.001) and the basal septum (9.8 ± 2.6 vs. 11.3 ± 1.7 cm/s, p=0.001).
  - S' values differ significantly between the groups in the basal septum (4.6 ± 1.7 vs. 5.7 ± 0.8 cm/s, p<0.001) but not in the LV lateral wall.
• At peak exercise:
  - LV lateral wall E' (13.2 ± 4.5 vs. 21.3 ± 2.5 cm/s, p<0.001) and S' (8.9 ± 5.4 vs. 14.2 ± 2.5 cm/s, p=0.001) velocities were significantly lower in HTx.
  - Also in the basal septum E' (9.7 ± 4.6 vs. 19.3 ± 2 cm/s, p<0.001) and S' (7.6 ± 3.2 vs. 11 ± 3.2 p=0.001) were lower in the HCM compared to controls.
  - The change in LV Lateral E' (5.7 ± 1.7 vs. 13.2 ± 4.7 p=0.02) and S' (3.4 ± 4 vs. 16.1 ± 4.8 p<0.01) and Septal E' (5.1 ± 2.9 vs 8.3 ± 2.2 cm/s, p<0.001) and S' (3.7 ± 2.3 vs 11.5 ± 6 cm/s, p=0.01) from baseline to peak was lower HCM compared to controls.
  - When looking at the TDI E' velocities and their relationship to increasing heart rate, HCM showed a lower intercept and lower values throughout all exercise stages, in both LV lateral wall and basal septum (Figure 2a and 2b).
  - Systolic TDI velocities showed the same velocity-frequency relationship pattern for the LV lateral wall and basal septum.
  - LV IVA values were significantly different at rest (1.22 ± 0.79 vs. 0.79 p<0.001) and at peak (2.4 ± 5.2 p<0.01) between HCM and controls.
  - The contractile response as studied by the FFR, was significantly blunted in HCM compared with controls, p=0.02. (Figure 3)

Conclusions
• In children with HCM, TDI LV systolic and diastolic velocities were reduced during exercise, as compared with healthy controls.
• The contractile response as studied by the FFR, was also significantly blunted in HCM children.
• The prognostic value needs further evaluation.