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**Prognostic significance of myocardial ischemia detected by single-photon emission computed tomography in children with hypertrophic cardiomyopathy.**

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Background: Myocardial ischemia caused by microvascular dysfunction is an important pathophysiologic component of hypertrophic cardiomyopathy (HCM), promoting myocardial fibrosis, adverse LV remodeling and impacting on clinical course and outcome in HCM patients. The aim of study was to assess the prevalence and clinical significance of myocardial ischemia in children with HCM using 99mTc-MIBI single-photon emission computed tomography (SPECT).
Methods: Seventy-nine children with HCM, median age 13.4yrs (IQR 9.9-15.5yrs), underwent SPECT evaluation from January 2006 to January 2015. Imaging were performed at rest and after maximal exercise.
Regional perfusion defects were identified in 59 of the 79 children (75%; gI), median age 13.6yrs (IQR 10.5-15.9yrs). Fixed perfusion defects were evident in 21 patients. In 38 children perfusion defects were present only during exercise and they completely resolved at rest. In 20 of the 79 children (25%; gII), median age 11.8yrs (IQR 7.9-15.2yrs) myocardial perfusion defects were not detected. Patients demographics, cardiovascular events, ECG, 24-h Holter ECG and myocardial fibrosis in CMR were analyzed and compared between the groups. The clinical endpoints were defined as cardiovascular events: sudden cardiac death (SCD), HF-related death, appropriate ICD discharges (ICDdx), resuscitated cardiac arrest (rCA), heart transplant (HTx), nonsustained ventricular tachycardia (NSVT), syncope, progression of heart failure to NYHA class III.
Results: During a median follow-up period of 6.1yrs (IQR 3.7-7.4yrs), in children with myocardial ischemia significantly more often clinical endpoints occurred (39 vs 6;p=0.048) and significantly more patients reached a clinical endpoint (28 [47%] vs 4 [20%];p=0.03).
In children with myocardial ischemia, the following cardiovascular events occurred: SCD (n=2), ICDdx (n=2), rCA (n=1), HF-related death (n=1), HTx (n=1), NSVT (n=12), syncope (n=11), progression to NYHA class III (n=9). In children with myocardial ischemia significantly more frequent myocardial fibrosis (68% vs 25%;p=0.019) was detected.
Conclusions: (1) Myocardial perfusion defects may reflect an ischemic process which affects the clinical manifestations in children with HCM. (2) Myocardial ischemia is an important predictor of adverse clinical events and risk of death in children with HCM. (3) Myocardial ischemia in HCM patients frequently correlates with myocardial fibrosis.