

Left ventricular diastolic dysfunction assessed by tissue Doppler imaging predicts clinical course in children with hypertrophic cardiomyopathy.

*Ziolkowska L., Boruc A., Kowalczyk M., Gorbacz-Mrowiec L., Brzezinska-Rajszyz G.
Department of Pediatric Cardiology
The Children's Memorial Health Institute, Warsaw, Poland*

Background: Tissue Doppler imaging (TDI) parameters have become a sensitive measure of left ventricular diastolic dysfunction (LVDD), which reflects the clinical course in children with hypertrophic cardiomyopathy (HCM). The aim of the study was to determine whether diastolic TDI parameters are predictive of adverse clinical outcome in children with HCM.

Methods: Sixty-three children, median age 14.3years with HCM studied since 2010 to 2014 were enrolled and prospectively followed with respect to TDI results and to clinical endpoints. Patients underwent echocardiography, ECG, Holter ECG, CMR-LGE. The LVDD was diagnosed if one or more parameters were raised in relation to the standards values: transmitral septal/lateral E/E' (z-score>2), LA dimension (z-score >2), LA volume index (>34 ml/m²). The clinical endpoints were defined as cardiovascular events: sudden cardiac death (SCD), appropriate ICD discharges (ICDdx), nonsustained ventricular tachycardia (NSVT), supraventricular tachycardia (SVT), syncope, progression of heart failure to NYHA class III. We analyzed also other parameters: chest pain, QTc, the presence of myocardial fibrosis in CMR-LGE. All 63 pts were divided into groups:gl-40(63%) children with LVDD and gII-23(37%) with normal LV diastolic function.

Results: During a follow-up, median 2.5years, 21(33%) children reached the clinical endpoints. In group with LVDD (gl) 19pts achieved endpoints: SCD(n=3), ICDdx(n=2), NSVT(n=3), SVT(n=2), syncope(n=2), progression to NYHA class III(n=7) while in gII without LVDD only 2pts: NSVT(n=1) and SVT(n=1); (p=0.0017).TDI parameters transmitral septal and lateral E/E' demonstrated increased values, furthermore septal E/E' ratio>2.21 z-score was proved to be an independent predictor of adverse clinical outcome. Moreover significantly larger LA dimension (median z-score 2vs0.7;p=0.02) and more often LA volume index>34 ml/m² (57% vs 26% pts;p=0.02) in children with clinical endpoints were found. In children with LVDD increased value of QTc (0.43vs0.41;p=0.032) and significantly more frequent myocardial fibrosis (33/63=52%vs5/63=8%;p=0.0001) were observed.

Conclusions: (1) TDI parameters are sensitive indicators of diastolic LV dysfunction, which is an important predictor of major clinical events and risk of death in children with HCM. (2) Measurement of transmitral septal E/E' should find a place in the routine clinical follow-up of children with HCM. (3) Myocardial fibrosis is highly significant predictor of LVDD in children with HCM.