Echocardiographic predictors of unfavourable outcome in children with hypertrophic cardiomyopathy

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Introduction

Echocardiographic parameters - the maximal wall thickness (MWT), left atrial dimension (LAD) and left ventricular outflow tract gradient (LVOTO) was reported to predict the cardiovascular events in adult patients with hypertrophic cardiomyopathy (HCM).

Aim

The aim of our study was to evaluate the progression of the disease expressed by these echocardiographic parameters and to determine whether they are predictive of adverse clinical outcome also in the pediatric population with HCM.

Methods

A total of 100 children with HCM aged 0-18 years (mean 9.4), examined since 2002 to 2017 were enrolled and prospectively followed with respect to echocardiography results and to clinical endpoints.

All children were classified as group I (gI, reached endpoint) or II (gII, not reached).

The echocardiographic parameters were collected from the first (H1) and last (H2) examination (time of observation 0.2-11.3 years, mean 3.1 years), and compared between groups I and II.

The clinical endpoints were defined as cardiovascular events: sudden cardiac death (SCD), heart failure cardiac death (HFCD), aborted cardiac arrest (CA), appropriate ICD discharges (ICDdx), heart transplantation (HTx).
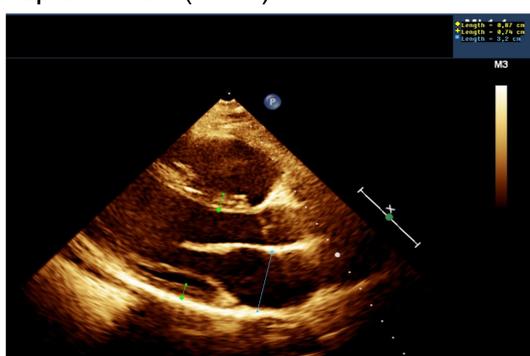

8-year-old girl: MWT (z-score 2.2) and LAD (z-score 5.4) during H1 (gI)

Results

During a follow-up, mean 7.3 years, 18(18%) of children reached the clinical endpoints (gI), while 82(82%) did not (gII). The following endpoints occurred in gI: SCD(n=3), HFCD(n=3), CA(n=2), ICDdx (n=6), HTx (n=4).

The difference in MWT progression was not significant between the two groups (z-score 2.9±4.6 vs 1.0±4.7; p=0.11), but significant progression of MWT (mean z-score from 10.8 [15.4mm] to 13.8 [19.9mm]; p=0.015) was observed in gI, while not in gII (mean z-score from 9.1 [14.9mm] to 10.1 [17.4mm]; p=0.07).

The same girl (13-year-old): MWT (z-score 19.5) and LAD (z-score 10.9) during H2 (gI)
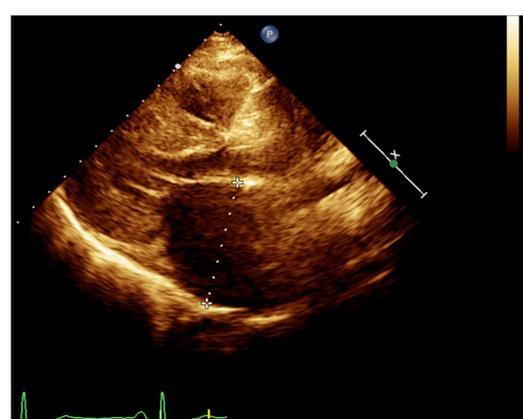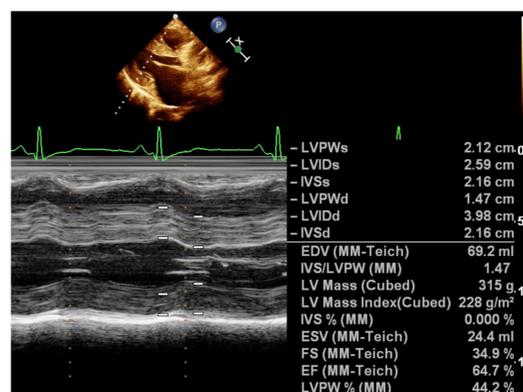
Results

LAD progression was significantly different between groups (z-score 1.1±3.0vs-1.3±2.4 respectively; p=0.001). LAD remained relatively stable enlarged in gI (z-score 4.1 [35.4mm] to 5.2 [39.7mm]; p=0.156) and decreased significantly in gII (z-score 3.6 [35.8mm] to 2.4 [24.8mm]; p<0.001). The progression of the LAD increased the odds of reaching the endpoint by 3.3 (p=0.04).

Progression of the disease and MWT (z-score 2.2 vs 19.5) and LAD (z-score 5.4 vs 10.9) in the same girl over 5 years of follow-up (gI)
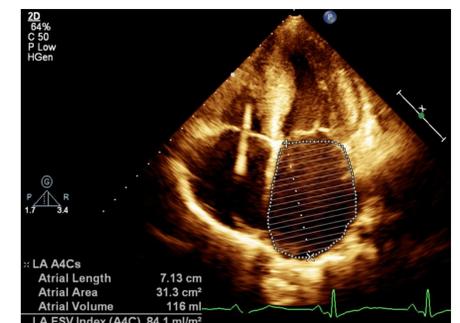
Changes in the value of echocardiographic parameters in the analyzed group of patients

Parameter	gI (n=18)	gII (n=82)	p
MWT-H1 (z-score)	10,8±6	9,1±6,4	0,3
MWT-H2 (z-score)	**13,8±6,1**	**10,1±7,0**	**0,039**
MWT progression	2,9±4,6	1,0±4,7	0,11
LAD-H1 (z-score)	4,1±4,3	3,6±3,0	0,57
LAD-H2 (z-score)	**5,2±4,8**	**2,4±2,8**	**0,001**
LAD progression	**1,1±3,0**	**-1,3±2,4**	**0,001**
LVOTO-H1 (mmHg)	20,0±23,1	18,7±25,2	0,85
LVOTO-H2 (mmHg)	24,0±28,7	21,7±29,6	0,77
LVOTO progression	4,0±13,8	2,9±18,7	0,82

LVOTO changes were insignificant in both groups, it increased from 21.4mmHg to 23.5mmHg and from 18.7mmHg to 21.7mmHg, respectively.

Conclusion

(1) Increase of MWT is a significant predictor associated with disease progression and unfavorable clinical outcome in pediatric population with hypertrophic cardiomyopathy.

(2) LAD progression is the most sensitive indicator of cardiovascular events in children with HCM, therefore should be measured in echocardiography regularly during follow-up.