

Reduced left ventricular end-diastolic volume in children with hypertrophic cardiomyopathy can be reproducibly measured with 3-D echocardiography and correlates with abnormal physiology

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BACKGROUND Hypertrophic cardiomyopathy (HCM) is associated with hypertrophy that reduces left-ventricular end-diastolic volume, often with an asymmetrical cavity that renders volume calculations by M-mode measurements invalid. The aim of this study was to investigate whether 3-D volume measurements of left ventricular (LV) volume can be used for assessing disease severity in children with HCM.

METHODS Ultrasound examinations were performed with Philips IE33 and analysed with 3-D QLab software (version 7). 80 healthy children and teenagers age 0.2 – 20 years were used to study the factors influencing LV volume, and 22 young HCM-patients (age 0.03 – 21 years; mean 10.4 years) who had had LV-volume measured before commencing therapy comprised the study group.

RESULTS LV volume in normal children correlated positively independently with both body surface area (BSA), and age. We also found a sizable gender difference in the 10-18 year olds, with males having 13% higher end-diastolic volumes (49.8 ± 5.3 versus 43.9 ± 4.5 ml/m² BSA; $p=0.0008$), and 17.6% higher stroke volumes (31.4 ± 3.2 versus 26.7 ± 3.1 ml/m² BSA; $p=0.00005$) than females. The smaller 0-9 year group showed a similar but non-significant trend. Each HCM-patient was given an age- and gender-matched control for statistical comparisons. The normal age-matched controls (average age 10.6 years) had a mean end-diastolic volume (EDV) of 44.9 ml/m² BSA [95% CI 41.2-48.6]; median 45.4 ml /m² BSA. HCM-patients had a mean EDV of 35.0 ml/m² BSA [30.3-39.7]; median 35.7 ml /m² BSA ($p=0.00019$ signed rank). Ejection fraction was higher in HCM-patients (median 69.2% versus 66.6% in controls; $p=0.0028$), but stroke volume at rest nevertheless remained lower in the HCM-patients (median 24.3 versus 28.3 ml/m² BSA; $p=0.00019$). Percent reduction in EDV correlated with diastolic dysfunction expressed as E:A ratio, and E:e ratio (correlation coefficients 0.44 and -0.47) and there was an inverse relationship with ejection fraction (-0.54). The reproducibility of EDV in HCM-patients between two independent observers was good with a mean difference of 0.17 ml/m² BSA [95% CI -0.7 till 1.0 ml/m² BSA].

CONCLUSIONS Reduced end-diastolic LV volume and stroke volume in children with hypertrophic cardiomyopathy correlates with abnormal physiology and may be a useful tool to assess disease progression.