Diffuse and Heterogenous Systolic Dysfunction in Hypertrophic Cardiomyopathy with Myocardial Tagging

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Background: Hypertrophic cardiomyopathy (HCM) is characterized by myocardial hypertrophy, which may be associated with myocyte disarray and fibrosis. These structural abnormalities may cause ventricular systolic and diastolic dysfunction. However, the relationship between structural and functional abnormalities has not yet been well defined. In a cohort of subjects with HCM, we studied the extent and pattern of left ventricular systolic impairment as assessed by tagged CMR in relation to wall thickness and delayed enhancement.

Materials & Methods: Thirty-eight patients (39.8±16.3 years, 26M) diagnosed with HCM based on clinical and echocardiographic assessment and 3 healthy controls (48 ±8.5yr, 2M) were enrolled in the study. CMR on a 1.5T, Philips MR scanner (Gyroscan ACS-NT, Philips Medical Systems, Best, NL) including non-tagged SSFP functional imaging, mid-ventricular short-axis tagged imaging using spiral complementary spatial modulation of magnetization (CSPAMM), and delayed enhancement imaging. Cine imaging with an SSFP sequence (Fig. 1A) and delayed enhancement imaging with a segmented inversion-recovery T1-TFE sequence (Fig. 1B) was performed at the same location as CSPAMM imaging. Minimal principal strain (**MPS**, a measure of primarily circumferential shortening with lower numbers indicative of greater shortening) between end-diastole and end-systole was calculated for 6 myocardial segments in endocardial, middle, and epicardial layers using a customized local-phase based algorithm (Ref. 1) implemented in MATLAB. To assess for functional differences in regions with and without hypertrophy, data were analyzed for the anterior junction region (AJ: including anterior septum and anterior segments, inferior junction region (IJ: inferior septal and inferior segments), and lateral free wall region (FW; anterolateral and inferolateral segments). Segments in HCM group were classified according to end-diastolic wall thickness (EDWT) in SSFP images with hypertrophied segments (HS) >= 12 mm of EDWT and non-hypertrophied segments < 12 mm (NHS). In addition, we analyzed MPS in relation to presence of delayed enhancement. Statistical analyses were performed with a p-value<0.05 considered significant.

Results: HCM group showed normal ejection fraction (69.4±8.02 %) and increased left ventricular mass index (99.56±26.95 g/m$_2$)(Ref. 2). MPS values stratified by epi-, mid-, and endocardial layers in the HCM group were significantly greater than those in the control group (-0.18±0.08 vs. –0.24±0.06; p=.001, -0.22±0.08 vs. –0.28±0.07; p=.001, -0.28±0.09 vs. –0.34±0.06; p=.001, respectively). MPS was significantly greater in epicardial layer of AJ and IJ (-0.16±0.07 vs. –0.22±0.05, p=0.02, -0.15±0.08 vs. –0.21±0.03, p=.002) (Fig. 2A), middle layer of IJ (-0.19±0.07 vs. –0.26±0.04, p=.004) (Fig. 2A), and endocardial layer of AJ (-0.28±0.10 vs. –0.37±0.05, p=.003) in the HCM group than in the control group. Linear Correlation coefficient (r) between MPS and EDWT was 0.43. HS (n=45) were found on anteroseptal (12), anterior (4), anterolateral (4), inferolateral (2), inferior (8), and inferoseptal walls (15). EDWT was significantly different between the control group, NHS, and HS (6.0±1.2 mm, 8.1±1.9 mm, 15.1±3.5 mm., respectively, ANOVA p<.05 in overall and between all groups). MPS values in each layer of myocardium were significantly different between the control group, NHS, and HS (epicardial; –0.24±0.06, -0.19±0.07, -0.13±0.07, middle; –0.29±0.07, -0.24±0.08, -0.15±0.06, endocardial; –0.35±0.06, -0.30±0.08, -0.22±0.08, ANOVA p<.05 in overall and between all groups except for between the control and NHS in endocardial layer). Delayed enhancement was found on 36 segments of 19 (50%) patients with HCM (anteroseptal 10, anterior 4, anterolateral 2, inferolateral 1, inferior 5, inferoseptal 14). EDWT of enhanced segments was significantly greater than that of nonenhanced segments (13.9±5.1 mm vs. 8.6±2.5 mm, p<.05). MPS values stratified by epi-, mid-, and endo-cardial layers in enhanced segments were significantly greater than those in the nonenhanced segments in HCM (epicarial; –0.13±0.06vs. –0.19±0.08; p=.001, mid; –0.16±0.06 vs. –0.24±0.08; p=.001, endocardial; –0.24±0.09 vs. –0.29±0.09, p=.001).

Summary: Systolic dysfunction in HCM is regionally heterogeneous and involves hypertrophied as well as nonhypertrophied segments. MPS is positively correlated with EDWT. The epicardial layer shows dysfunction more frequently than mid or endocardial layer. Tagged MRI may be helpful for detection of transmural and nontransmural dysfunction in nonhypertrophied and hypertrophied segments. Delayed enhancement was frequently found in septal junction regions and in segments with marked hypertrophy, which showed more systolic dysfunction.

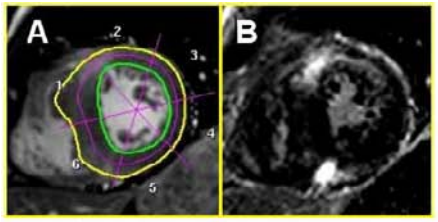

Fig. 1. (above) (A) End-diastolic cine SSFP image with prominent septal hypertrophy, (B) corresponding delayed enhancement image with delayed hyperenhancement at AJ and IJ.

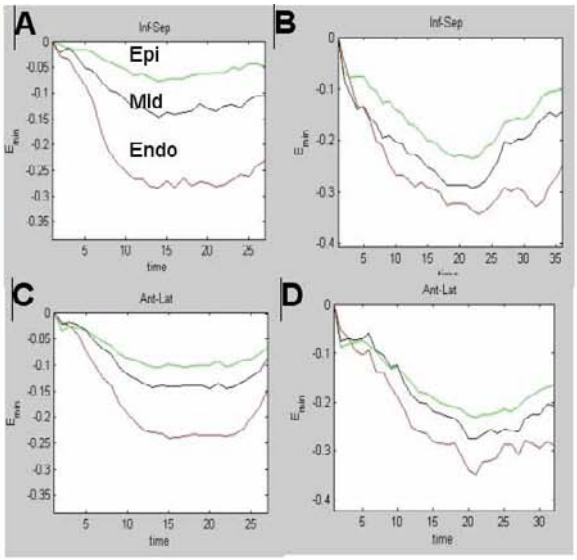

Fig. 2. (to right) (A,C) MPS in inferior septum (#6 in Fig. 1A) and anterolateral wall (#3 in Fig. 1A) of subject in Figure 1(A). (B,D), Corresponding MPS in a control. MPS in epi- and mid-layer showed a marked increase compared to that in the control,not only on the hypertrophied inferior septum (17.3mm) but also on normal-thick anterolateral wall (10.1mm).

References:

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