Differences in Global and Regional Left Ventricular Myocardial Mechanics in Various Morphologic Subtypes of Patients With Obstructive Hypertrophic Cardiomyopathy Referred for Ventricular Septal Myotomy/Myectomy

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Patients with obstructive hypertrophic cardiomyopathy (HC) have various left ventricular (LV) shapes: reverse septal curvature (RSC, commonly familial), sigmoid septum (SS, common in hypertensives), and concentric hypertrophy (CH). Longitudinal (systolic and early diastolic) strain rate (SR) is sensitive in detecting regional myocardial dysfunction. We sought to determine differences in longitudinal SR of patients with obstructive HC, based on LV shapes. We studied 199 consecutive patients with HC (50% men) referred for surgical myectomy. Clinical and echocardiographic parameters were recorded. LV shapes were classified on echocardiography, using basal septal 1/3 to posterior wall ratio: RSC = ratio >1.3 (extending to mid and distal septum), SS = ratio >1.3 (extending only to basal 1/3), and concentric = ratio ≤1.3. Longitudinal systolic and early diastolic SRs were measured from apical 4- and 2-chamber views (VVI 2.0; Siemens, Erlangen). Distribution of RSC, SS, and CH was 50%, 28%, and 22%, respectively. Patients with RSC were significantly younger (47 ± 12 vs 64 ± 10 and 57 ± 11, respectively) with lower hypertension (40% vs71% and 67%, respectively) than patients with SS or CH (both p <0.001). Patients with RSC had lower global systolic (−0.99 ± 0.3 vs −1.05 ± 0.3 and −1.17 ± 0.3) and early diastolic SR (0.95 ± 0.4 vs 0.98 ± 0.3 and 1.16 ± 0.4) versus patients with SS and CH (in 1/s, both p <0.01), despite being much younger and less hypertensive. RSC was associated with abnormal global LV systolic (beta 0.16) and early diastolic (beta −0.17) SR (both p <0.01). In conclusion, patients with HC with RCS have significantly abnormal LV mechanics, despite being younger and less hypertensive. A combination of LV mechanics and shapes could help differentiate between genetically mediated and other causes of obstructive HC. **(Am J Cardiol 2014;113:1879−1885)**

A variety of echocardiography-based techniques have been utilized to assess global and regional left ventricular (LV) mechanics (strain and strain rate [SR]), in patients with hypertrophic cardiomyopathy (HC) and various other disorders.[1−5] Although the phenotypic expression of hypertrophy, obstruction, and diastolic dysfunction are typically progressive in HC, there is increasing recognition that there are changes in regional LV function (both systolic and diastolic) that predate overt morphologic and functional LV changes.[1,2] Recently, speckle tracking echocardiography (STE) has emerged as an accurate and sensitive tool to study global and regional LV mechanics.[6,7] There are no data describing differences in LV myocardial mechanics and deformation, based on the morphologic shape of the LV and ventricular septum, in patients with HC. We hypothesized that patients with reverse septal curvature (RSC, more likely to represent genetically mediated form of HC) would have worse global and regional LV mechanics, despite having similar degree of LV outflow obstruction, compared with sigmoid septum (SS) and concentric hypertrophy (CH; other morphologic subtypes seen in HC). We sought associations of systolic and diastolic LV mechanics, measured by STE, with the different morphologic shapes in adult patients with HC with similar degree of symptomatic LV outflow obstruction who underwent surgical myectomy.

Methods

The study population consisted of 199 consecutive patients with HC with severe LV outflow obstruction who underwent surgical myectomy at our tertiary referral center. We excluded patients with fixed obstruction (subaortic membrane or aortic stenosis) that also had a concomitant myectomy, as these patients have a different pathophysiologic profile. To maintain homogeneity of the study population, in terms of afterload, LV outflow obstruction, and gradients, we only selected patients with severe LV outflow obstruction who underwent surgical myectomy. An initial clinical diagnosis of HC was established after a thorough

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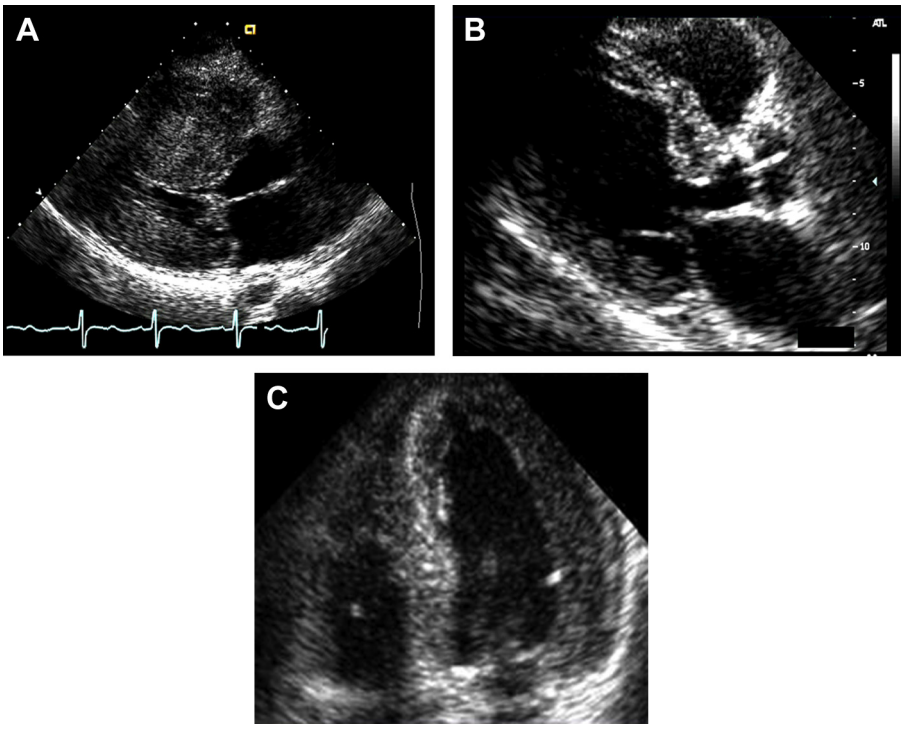

Figure 1. *(A)* Parasternal long-axis echocardiographic image in a patient with RSC. Notice the predominant midseptal convexity toward the LV cavity, with the cavity having a crescentic shape. *(B)* Parasternal long-axis echocardiographic image in an older hypertensive patient with sigmoid septum. Notice the ovoid cavity with the septum being concave toward the LV with a pronounced basal septal bulge, and *(C)* 4-chamber echocardiographic image in a patient with CH, where there was a similar degree of hypertrophy throughout the LV. All patients had severe resting or provokable LV outflow tract gradient, requiring surgical myectomy.

history, examination, electrocardiography, and imaging. HC was defined as a hypertrophied and nondilated left ventricle in the absence of another cardiac or systemic disease that could result in a similar magnitude of hypertrophy.[8] All patients had symptoms attributable to severe dynamic LV outflow obstruction, despite optimal medical therapy at maximum tolerated dosages and were referred for surgical intervention after consensus between cardiologists and cardiothoracic surgeons. Baseline demographic, clinical, and imaging data were collected. The study patients are part of an institutional review board−approved registry.

All patients underwent comprehensive echocardiograms using commercially available instruments (HDI 5000; Philips Medical Systems, N.A., Bothell, Wash and Acuson Sequoia; Siemens Medical Solution USA, Inc., Malvern, Pennsylvania) as part of standard workup. End-diastolic interventricular septal and posterior wall thickness was measured in a standard fashion according to the guidelines.[9] LV ejection fraction and volumes were measured, using the standard short-axis, 2-, and 4-chamber views.[9] LV volumes were subsequently indexed to body surface area. Resting LV outflow peak velocity was measured by a continuous-wave Doppler echocardiography, and resting LV outflow pressure gradient was estimated using simplified Bernoulli equation.[10] Care was taken to avoid contamination of the LV outflow waveform by mitral regurgitation jet. In patients with resting left ventricular outflow tract gradients <30 mm Hg, provocative maneuvers, including Valsalva, amyl nitrite, and exercise echo, were also used to measure a provocable LV outflow gradient. In patients with resting peak LV outflow gradient >100 mm Hg, provocative

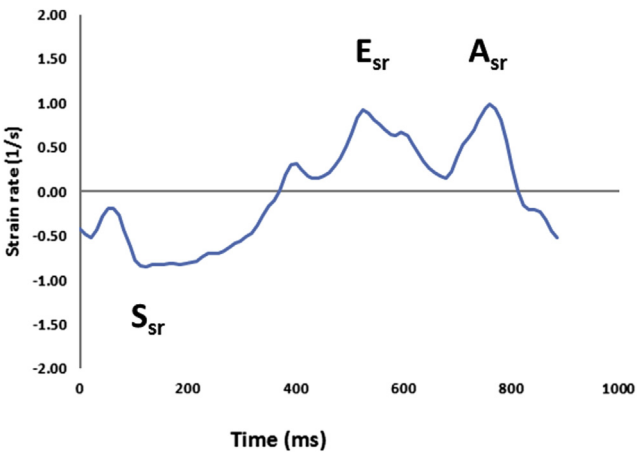

Figure 2. A representative image showing measurement of global longitudinal SR in a patient with HC. Only systolic and early diastolic SR measurements were analyzed in the study. Ssr = systolic SR; Esr = early diastolic SR; Asr = late diastolic SR.

maneuvers were not used. Maximal LV outflow gradient was recorded and defined as the highest recorded gradient (resting or provoked) in a given patient.[11] In addition, resting afterload was roughly estimated as the sum of resting LV outflow gradient + systolic blood pressure (mm Hg). Degree of resting mitral regurgitation was assessed by color Doppler and quantified according to the multiple established criteria, on a scale of 0 to 4+ (0 = none, 1+ = mild, 2+ = moderate, 3 = moderately