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Quantitation of Localized Abnormal Deformation in Asymmetric Nonobstructive Hypertrophic Cardiomyopathy: A Velocity, Strain Rate, and Strain Doppler Myocardial Imaging Study

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Abstract. We report a case of a 10-year-old child with nonobstructive hypertrophic cardiomyopathy in whom two-dimensional echocardiography showed asymmetric septal hypertrophy with a localized thickening in the mid-septal segment. Systolic regional longitudinal motion and deformation indices were quantified by the new ultrasound-based parameters velocity, strain rate, and strain. Regional longitudinal myocardial function indices were normal for the basal and apical septal segments. The deformation parameters strain rate and strain (not the regional velocity profile) were abnormal only in the hypertrophied mid-septal segment with myofibril disarray. The concepts and advantages and clinical implications behind this quantitative approach to localizing and quantifying areas of abnormal deformation related to such myocardial disarray in localized hypertrophy are discussed.

Key words: Regional myocardial function — Hypertrophic cardiomyopathy — Echocardiography

Children with hypertrophic cardiomyopathy frequently present with localized hypertrophic segments [10]. Myocardial fiber disarray is the usual histologic finding in such hypertrophic segments.

Echocardiography is a well-established clinical tool for the noninvasive assessment of both regional and global left ventricular function in children with hypertrophic cardiomyopathy [11]. In pediatric cardiology, the quantification of myocardial function in hypertrophic segments is most often based on either visual interpretation of wall motion or on conventional M-mode echocardiography. However, the visual interpretation of either myocardial motion or local myocardial deformation is nonquantitative, subjective, and experience dependent [5]. On the other hand, quantitative measurement of radial function from the parasternal views is limited to only a few segments. In contrast, longitudinal function measurement from the apical segments by conventional M-mode echocardiography for ring displacement only quantifies the overall shortening or lengthening of the whole subtended myocardial wall and does not measure the deformation in the segment with local hypertrophy.

Doppler myocardial velocity (VEL) measurement has been shown to provide a new method of quantifying regional myocardial motion in children [6]. However, because myocardial velocities detect only regional motion rather than myocardial wall deformation, motion in one segment may be influenced by motion induced to tethering of adjacent myocardial segments [4]. Ultrasonic strain rate (SR) and strain (ε) measurements provide a new noninvasive method of quantifying local changes in myocardial deformation [2] which are relatively independent of the deformation in adjacent segments. SR (i.e., the rate of deformation) is a measure of the regional spatial gradient in myocardial velocities and represents the local velocity of deformation [1]. Regional ε is the time integral of SR and represents the local magnitude of deformation expressed as a percentage [1].

This new noninvasive quantitative approach could be of value in determining the site and extent of myocardial fiber disarray and the regional myocardial function in the different hypertrophic and nonhypertrophic segments.

Methodology

Real-time two-dimensional (2D) color Doppler myocardial imaging (CDMI) data were recorded from the left ventricle using standard apical views at a high frame rate of 150 ± 10 frames/sec (GE Vingmed System V, Horten, Norway; 3.5 MHz). An

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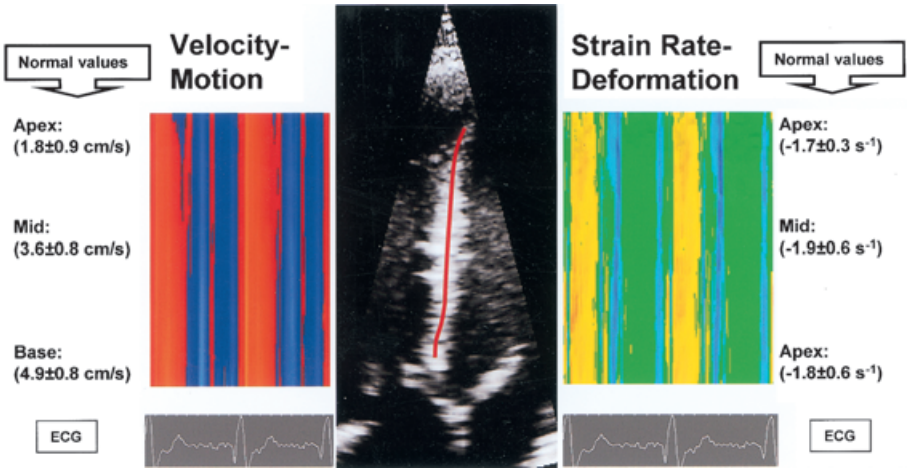

Fig. 1. The assessment of regional septal longitudinal myocardial function in a normal child. This was assessed using color-coded longitudinal velocity (*VEL*) and strain rate (*SR*) curved M-modes computed from color Doppler myocardial imaging data obtained by scanning the long-axis view from the apex. (***Left***) A color-coded VEL map displayed in the curved M-mode format with regional velocities displayed from base to apex. The red color in systole indicates motion toward the transducer and the blue color in diastole indicates motion away from the transducer. (***Middle***) An apical long-axis view with the septum in the middle of the scan sector. The red line indicates the position of the interrogating curved M-mode. (***Right***) The color-coded SRs from base to apex displayed in the curved M-mode format. The orange color in systole indicates myocardial shortening and the blue color in diastole indicates lengthening.

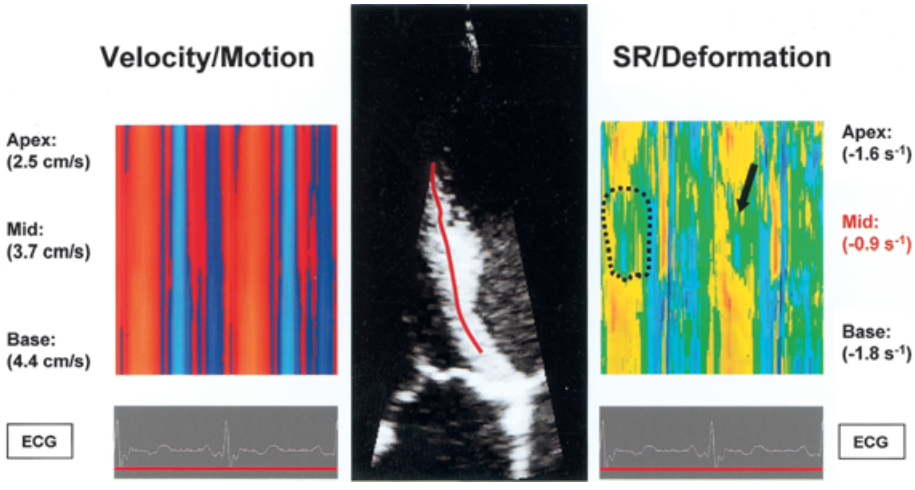

Fig. 2. The assessment of regional septal longitudinal myocardial function in a patient with localized hypertrophy. This was assessed using color-coded longitudinal velocity (*VEL*) and strain rate (*SR*) curved M-modes computed from color Doppler myocardial imaging data obtained by scanning the long-axis view from the apex. (***Left***) A color-coded VEL map displayed in the curved M-mode format with regional velocities displayed from base to apex. The red color in systole indicates motion toward the transducer and the blue color in diastole indicates motion away from the transducer. (***Middle***) An apical long-axis view with the septum in the middle of the scan sector. Notice the localized hypertrophy in the mid-septal segment. The red line indicates the position of the interrogating curved M-mode. (***Right***) The color-coded SRs from base to apex displayed in the curved M-mode format. The apical and basal segments shorten in systole (*orange band*) and lengthen in early and late diastole (*blue band*). In contrast, almost no deformation is present in the mid-septum in systole (*green band indicated by arrow and circumferenced area*).

appropriate velocity scale was chosen in order to avoid CDMI data aliasing. The narrowest image sector angle possible (usually 30°) was used to achieve the maximum color Doppler frame rate possible. Care was taken to keep the septum in the center of the ultrasound sector so that it was aligned at as near zero degrees as possible to longitudinal motion.

Off-Line Analysis

CDMI data were stored in digital format and transferred to a computer workstation for off-line analysis using dedicated software (TVI, GE Vingmed). This allowed the computation of regional VEL, SR, and ε values [2].

VEL. Regional VELs were extracted from CDMI clips. According to convention, longitudinal motion in systole directed toward the transducer is ascribed positive values and color coded red to yellow, and motion in diastole away from the transducer is ascribed negative values and coded blue to green (Fig. 1).

SR. This measures the local rate of deformation of a tissue. If derived from a myocardial wall using ultrasound velocity measurements, it corresponds to the local spatial velocity gradient and it can be expressed in sec^{-1}. For the longitudinal direction when the segment shortens (systole), it is given a negative SR value and is color coded yellow to red. When the segment lengthens (diastole), it is characterized by a positive value and is coded from cyan to blue. In addition, SR values near zero are colored green (Fig. 1).

ε. This defines the amount of local deformation in terms of percentage and is derived by the integration of the SR curves. Myocardial longitudinal ε values describe regional shortening in systole and are expressed as negative values, whereas any regional lengthening in diastole is expressed as a positive value. Analysis was performed for regional longitudinal peak systolic VEL and peak systolic SR and systolic ε for the basal, mid-, and apical segments of the septum.

Case Report

A 10-year-old asymptomatic boy with localized nonobstructive hypertrophic cardiomyopathy was studied. The 12-lead electrocardiogram (ECG) and chest x-ray were both normal. Standard gray scale and blood pool Doppler ultrasound examinations revealed the typical ultrasound findings diagnostic of a localized hypertrophic cardiomyopathy involving the mid-segment of the interventricular septum (end diastolic wall thickness 14.7 mm). There was no evidence of left ventricular outflow tract obstruction (peak velocity in the outflow tract 1.3 m/sec) at rest, but there was mild (2/4) mitral insufficiency (color jet reaches first third of the left atrial cavity; normal size of the left atrium 29 mm). The basal and apical segments of the septum were within the normal range of wall thickness measurements (end diastolic wall thickness: basal 7 mm; apical 8 mm). Left ventricular dimensions (LV end diastolic diameter = 3.45 cm) and global function were normal (LV ejection fraction = 60%).

Following the conventional echocardiographic studies, CDMI data were recorded from the left ventricular septum using an apical view. Data were acquired at a high frame rate of 150 frames/second (GE Vingmed System V; 3.5 MHz) and transferred to a computer workstation for off-line analysis using dedicated software which allowed the computation of regional VEL, SR, and ε values [3]. Off-line analysis was performed to determine the regional longitudinal peak systolic VEL, peak systolic SR, and systolic ε for the basal, mid-, and apical segments of the septum using the methodology previously described [3].

The color-coded longitudinal (i.e., base–apex) velocities of septal motion (cm/sec) derived by curved M-mode showed homogeneous systolic motion toward the apical transducer position (Fig. 2, red band) and early and late diastolic motions away from the transducer (blue bands). The corresponding curved M-mode for regional longitudinal SR (sec^{-1}) revealed shortening in systole in the apical and basal segments (orange band) with a normal lengthening pattern in early and late diastole (blue band). In contrast, almost no systolic deformation was present in the abnormally hypertrophied mid-septal segment (Fig. 2, green band indicated with arrow).

The regional myocardial peak systolic velocities were in the normal range for all three segments. Regional myocardial peak systolic SRs were normal for the basal and apical segments (SR base = -1.8 sec^{-1}; SR apex = -1.6 sec^{-1}) but reduced for the midsegment (SR = -0.9 sec^{-1}). Regional changes in ε paralleled the SR changes. Regional myocardial systolic ε values were normal for the basal and apical segments (ε basal = -19%; ε apical = -23%) but reduced for the midsegment (ε = -9%) (Fig. 3). As reference normal values, we used our VEL, SR, and ε data values obtained from 30 normal children [9].

Discussion

Noninvasive quantification of regional myocardial deformation properties by ultrasonic SR and ε has made it possible to study the relation between regional morphological abnormalities in hypertrophied ventricles and the longitudinal function abnormalities in the hypertrophied region. In this study, we have shown that the regional longitudinal deformation parameters SR and ε can differentiate abnormal deformation in a hypertrophic segment from normal deformation in nonhypertrophic segments. Our findings are consistent with the functional changes previously described by Wigle et al. [10]. In our patient, using ultrasound-based quantitation of deformation properties it was possible to localize the extent of impaired myocardial function within the wall.

The fact that the VEL measurements in the three segments were normal shows how tethering of adjacent segments (in this case the apical segment) may influence the measurement of regional motion (VEL) but not the measurements of regional deformation (SR and ε). This is in agreement with the study of Urheim et al. [7], who validated ε measurements with sonomicrometry and suggested that this deformation parameter may be less influenced by tethering effects than regional velocity estimation and thus represents true local deformation changes. In pediatric cardiology these new ultrasound indices could be important as a noninvasive approach for characterization of local myocardial deformation in acquired or congenital heart disease. The postprocessing algorithms allow data to be postprocessed in a matter of minutes after real-time data acquisition. Normal systolic and diastolic SR and ε values from all segments of the left and right ventricles in healthy children have already been defined [9]. This study showed that the left ventricular longitudinal SR and ε were homogeneous for left ventricular basal, mid-, and apical segments. On the other hand, longitudinal SR and ε values were significantly higher and heterogeneous in the right ventricle (compared to left ventricular walls) and were maximal in the mid-part of the right ventricular free wall (peak systolic SR, -2.8 ± 0.7 sec^{-1}; systolic ε, $-45 \pm 13\%$).

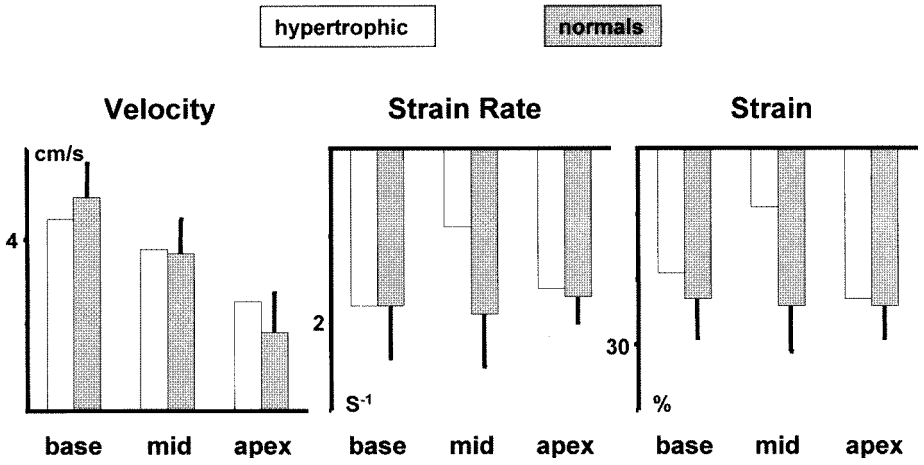

Fig. 3. A comparison of myocardial motion (VEL) and deformation parameters (SR/ε) for normal subjects versus the patient with localized hypertrophy. Note that the peak systolic velocity is in the normal range for the three septal segments (*left*). In contrast, regional peak systolic SR (*middle*) and systolic ε (*right*) are markedly reduced in the mid-septal hypertrophic segment.

In addition, SR and ε measurements have been shown to identify and quantify regional function in children with a systemic right ventricle after a Senning procedure [8]. In post-Senning patients, right ventricular regional systolic and diastolic longitudinal function indices were reduced compared to SR/ε values in low-pressure right ventricles. Moreover, the SR and ε values within the right ventricular free wall were very homogeneous, as in the normal systemic left ventricular free wall.

Despite these encouraging results, additional clinical studies are necessary to determine the precise clinical role of ultrasound-based deformation imaging in quantifying regional function in pediatric cardiology.

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References

1. Dhooge J, Heimdal A, Jamal F, et al (2000) Regional strain and strain rate measurements by cardiac ultrasound: principles, implementation and limitations. *Eur J Echocardiogr 1*:154–170
2. Heimdal A, Stoylen A, Torp H, Skjaerpe T (1998) Real-time strain rate imaging of the left ventricle by ultrasound. *J Am Soc Echocardiogr 11*:1013–1019
3. Jamal F, Kukulski T, Strotmann J, et al (2001) Quantitation of the spectrum of changes in regional myocardial function during acute ischaemia in closed-chest pigs. An ultrasonic strain rate and strain study. *J Am Soc Echocardiogr*, in press
4. Miyatake K, Yamagisi M, Tanaka N, et al (1995) New method for evaluating left ventricular wall motion by color-coded tissue Doppler imaging: *in vitro* and *in vivo* studies. *J Am Coll Cardiol 25*:717–724
5. Picano E, Lattanzi F, Orlandini A, Marini C, L'Abbate A (1991) Stress echocardiography and the human factor: the importance of being expert. *J Am Coll Cardiol 17*:666–669
6. Rychik J, Tian ZY (1996) Quantitative assessment of myocardial tissue velocities in normal children with Doppler tissue imaging. *Am J Cardiol 77*:1254–1257
7. Urheim S, Edvardsen T, Torp T, Angelsen B, Smiseth O (2000) Myocardial strain by Doppler echocardiography. Validation of a new method to quantify regional myocardial function. *Circulation 102*:1158–1164
8. Weidemann F, Eyskens B, Jamal F, et al (2000) Regional strain distribution in the right ventricle after Senning repair for d-transposition of the great arteries. A colour Doppler myocardial imaging study. *Eur Heart J 21*(Suppl): 616 [abstract]
9. Weidemann F, Jamal F, Mertens L, et al (2000) Regional strain distribution throughout the left and right ventricle in healthy children. A colour Doppler myocardial imaging study. A new cardiac ultrasound index of regional deformation. *Circulation 102*(Suppl 2):772 [abstract]
10. Wigle ED, Rakowski H, Kimball BP, Williams WG (1995) Hypertrophic cardiomyopathy. Clinical spectrum and treatment. *Circulation 92*:1680–1692
11. Yetman AT, Hamilton RM, Benson LN, McCrindle BW (1998) Long-term outcome and prognostic determinants in children with hypertrophic cardiomyopathy. *J Am Coll Cardiol 32*:1943–1950