CASE REPORTS

Utility of tissue characterization in apical hypertrophic cardiomyopathy diagnosis

João Abecasis*, Raquel Dourado, Isabel Arroja, José Azevedo, and Aniceto Silva

Cardiologia, Centro Hospitalar de Lisboa Ocidental, Rua Professor Reynaldo dos Santos, 2795-523 Carnaxide, Portugal

Received 16 March 2008; accepted after revision 8 August 2008; online publish-ahead-of-print 27 August 2008

A 60-year-old male with previous hypertension, left ventricle hypertrophy, and coronary artery disease was referred for stress echocardiography because of exertional chest pain. The electrocardiogram revealed deep T-wave inversion in the anterolateral leads. Contrast echocardiography was notable for an apical filling defect consistent with the apical form of hypertrophic cardiomyopathy. Cardiac magnetic resonance demonstrated the ‘ace of spades’ left ventricle cavity, confirming the diagnosis. Single photon emission computed tomography showed increased apical left ventricle tracer uptake. Velocity vector imaging study depicted lower than normal absolute maximal longitudinal tissue velocities. The apical longitudinal strain was negative without base to apex gradient. There were normal longitudinal strain values in the basal and mid myocardial segments (Figure 1). Apical hypertrophic cardiomyopathy is a rare condition occasionally missed by conventional echocardiographic studies. Intravenous contrast enhancement might improve diagnosis accuracy. Newer Doppler-based techniques allowing tissue characterization may complement contrast echocardiography in its diagnosis.

KEYWORDS
Apical hypertrophic cardiomyopathy; Velocity vector imaging; Tissue characterization

Case report

A 60-year-old Caucasian male was referred for stress echocardiography because of exertional chest pain. He had past history of hypertension and coronary artery disease with left anterior descendent artery percutaneous intervention. Mild left ventricle concentric hypertrophy was documented in a previous two-dimensional (2D) transthoracic echocardiographic study.

Cardiovascular exam was unremarkable and electrocardiogram revealed deep T-wave inversion in the anterolateral leads. When performing echocardiographic stress protocol with ultrasonic contrast enhancement (Sonovue®), an apical filling defect with a spade-like configuration consistent with apical hypertrophic cardiomyopathy was revealed (Figure 1, Supplementary material online, Clip S1). The apical thickness was 24.9 mm, the ratio of apical thickness to posterior wall thickness was >1.5, and the left ventricle mass was 250.5 g/m². The left ventricle systolic function was normal and the lateral mitral annular velocity was attenuated at 8 cm/s.

Cardiac magnetic resonance demonstrated the ‘ace of spades’ left ventricle cavity, confirming the diagnosis of apical form of hypertrophic cardiomyopathy (Figure 1). Single photon emission computed tomography was notable for increased apical left ventricle tracer uptake without perfusion defects in the stress study (Figure 2).

Velocity vector imaging (VVI) study—Siemens Syngo US workplace, left ventricle endocardium contour—is a new tracking algorithm technique applied to DICOM format 2D images, which is independent of ultrasound beam alignment or transducer location. It uses multiple tracking techniques to determine cardiac motion, including speckle tracking, mitral annulus displacement, and blood-tissue interface. At each stage of the tracking, Fourier analysis is used and applies the constraint that the trace returns to the same location at the subsequent cardiac cycle. This provides global and regional information concerning myocardial velocities, strain, strain rate, and synchrony.

In this case, the left ventricle VVI analysis was performed in apical four-chamber view and because of endocardium restricted analysis from the available software version (1.0), only endocardial border tracing was performed. The analysis depicted lower than the normal absolute maximal longitudinal tissue velocities, as was previously known from lateral mitral annular velocity by tissue Doppler imaging (TDI). There was a normal base to apex decreased

* Corresponding author. Tel: +351914054977 (J.A.)/+351914952818 (R.D.); fax: +351214241388
E-mail address: joaoabecasis@hotmail.com (J.A.)/raqueldourado@yahoo.com (R.D.).

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2008.
For permissions please email: journals.permissions@oxfordjournals.org.

doi:10.1093/ejechocard/jen227
There were normal longitudinal strain values in the basal and mid-myocardial segments, previously identified as hypertrophic segments by conventional 2D echo. However, the apical longitudinal strain was negative without base to apex gradient. When time to peak tangential velocity was assessed at six different left ventricle points, only the left apical segment was non-significantly delayed (32 s). All other segments simultaneously reached peak velocities and
this could be confirmed by the $< 10\%$ shift of the sinusoid of each segment (time to peak velocity curve Fourier analysis—phase) towards the average global phase (Figures 1 and 4).

**Discussion**

The apical form of hypertrophic cardiomyopathy is a rare condition initially described in Japanese population. Giant negative T waves in precordial leads are common electrocardiographic findings raising diagnosis suspicion. This condition can be missed by regular echocardiography, and traditionally definitive diagnosis has been made by left ventriculography demonstrating a spade-like appearance.\textsuperscript{1,2} Magnetic resonance imaging has also been used to establish the diagnosis obviating the need for invasive procedures.\textsuperscript{3}

Echocardiographic image enhancement with an intravenous contrast agent has been shown to improve diagnosis accuracy. Endocardial-blood interface may be better visualized and delineation of the apical border permits regional myocardial thickness measurement.
Newer Doppler-based techniques allowing tissue characterization and preclinical myocardial dysfunction evaluation may also improve diagnosis assessment.4–7 Tissue Doppler imaging provides information concerning regional basal and mid left ventricle segments behaviour. However and because of insonation angle dependency, left ventricular apex remains inaccessible to TDI study. Two-dimensional strain techniques, both by speckle tracking imaging and by VVI analysis, assess deformation by multiple tracking techniques (‘myocardial feature tracking’) being unique for left ventricle apical study.

Hypertrophic cardiomyopathy is classically associated with myocardial relaxation and filling abnormalities despite normal left ventricle ejection fraction.5 Regional systolic function may also be impaired in preclinical states and this could be evaluated by 2D strain imaging.

In spite of recently reported paradoxical longitudinal strain in the apical segments of apical hypertrophic cardiomyopathy patients studied by speckle tracking imaging 2D strain,8 we could not confirm this abnormality. However, when VVI tissue characterization was performed, we found abnormal regional velocities and deformation parameters, particularly concerning base to apex longitudinal strain gradient. As previously reported, this could be related to the abnormal tissue hypertrophy extending beyond the more evident apical hypertrophic segments.

Conventional echocardiographic techniques can misdiagnose apical hypertrophic cardiomyopathy. Apical endocardial border may even remain inadequately defined with second harmonic imaging, and contrast enhancement could obviate this fact permitting accurate thickness measurement. Left ventricle apex tissue characterization cannot be made by TDI albeit 2D strain by VVI might have a potential role in this assessment.

In spite of this, VVI needs further clinical validation, particularly regarding comparison with 2D strain analysis by similar techniques such as that provided by speckle tracking. It should also be noted that this software vectorial analysis was restricted to endocardial border assessment. In this case, the endo-epicardial myocardial deformation gradient9 could not be evaluated and this might be related to different speckle tracking findings in other case reports.

Multiple echocardiographic modalities might be useful in apical hypertrophic cardiomyopathy diagnosis. Newer 2D tissue characterization techniques appear to complement routine and contrast echo derived diagnostic information, probably obviating the need for other, possible more expensive and invasive, imaging studies.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

References