CASE REPORTS

Apical hypertrophic cardiomyopathy: potential utility of Strain imaging

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Hypertrophic cardiomyopathy (HCM) is a frequently recognized condition on echocardiography. The apical variant, also known as ‘Japanese variant’, is rare and often poses a diagnostic challenge. There has been a resurgence of interest in the diagnosis of HCM especially with the advent of novel imaging modalities such as strain imaging. Two-dimensional (2D) strain echocardiography calculates tissue velocities via frame-to-frame tracking of acoustic markers within the image and provides strain parameters comparable with tissue-Doppler-derived strain. We describe paradoxical apical strain (systolic lengthening) without overt apical dyskinesis (conventional imaging) in two patients with apical HCM, using 2D strain imaging. Our report highlights a novel application of 2D strain imaging that could facilitate the diagnosis of apical HCM.

KEYWORDS
Apical hypertrophic cardiomyopathy; Speckle tracking; Strain imaging

Introduction

Hypertrophic cardiomyopathy (HCM) is a frequently recognized entity on echocardiography. There has been a resurgence of interest in this entity, especially with the advent of new Doppler-based techniques such as strain imaging. However, Doppler-only based techniques are limited because of angle dependence of the signal precluding the assessment of apical left ventricular function. The apical variant of HCM, also known as ‘Japanese variant’, is encountered rarely but often presents a diagnostic challenge. We describe paradoxical apical longitudinal strain (systolic lengthening) in two patients with apical HCM in the absence of overt regional wall motion abnormalities using two-dimensional (2D) strain imaging, also known as speckle tracking imaging. This novel technology is not Doppler angle-dependent and might have a potential role in the rapid diagnosis of this condition.

Patient characteristics and imaging

Case 1

A 47-year-old Caucasian male with history of dementia secondary to leukodystrophy was referred for a 2D echocardiogram for evaluation of recurrent strokes. He had no symptoms suggestive of arrhythmias, congestive heart failure, or coronary artery disease. Cardiovascular exam was unremarkable, electrocardiogram revealed sinus bradycardia, first degree AV block, and signs of left ventricular hypertrophy (LVH) with deep T-wave inversion in the anterolateral leads.

Echocardiography showed normal left ventricle (LV) size and ejection fraction. The salient finding was the presence of severe LVH at the apex, measuring 29 mm in thickness. The basal portion of the LV wall was mildly hypertrophied with a wall thickness of 15 mm. No intracavitary gradient was recorded on Doppler interrogation. The left atrium was mildly dilated and right heart chambers were within normal limits. Obliteration of the LV apical cavity in systole without apparent regional wall motion abnormalities was observed (Figure 1). Myocardial contrast echocardiography showed preserved perfusion in the apical segments (Figure 2).

Two-dimensional strain analysis performed offline using a dedicated software package (EchoPac—GEVingMed, Milwaukee, WI, USA) showed gross attenuation of average longitudinal strain in the mid-myocardial segments compared with the basal myocardial segments (−12 vs. −21%, respectively) and paradoxical longitudinal strain (systolic lengthening) in the apical segments (+9.2%, Figure 3), supporting the diagnoses of apical variant of HCM. Typically, apical strain values in normal individuals are negative (−23%), reflecting maximal systolic shortening, whereas normal strain values in the basal and mid-myocardial segments have been reported to be in the range of −16.0 to −19.0%, respectively, with a progressive increase in longitudinal strain from base to apex (base–apex gradient).1,2

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Case 2

A 26-year-old otherwise healthy Caucasian male was evaluated for recurrent syncopal spells with exertion. He denied exertional chest pain or shortness of breath. No past history of hypertension, diabetes, valvular, or congenital heart disease. Physical exam was unremarkable. A 12-lead ECG showed increased QRS voltages suggestive of LVH and deep, symmetrically inverted T waves without evidence of preexcitation. A surface echocardiogram was notable for normal left ventricular systolic function with asymmetric hypertrophy of the apical segments. Myocardial thickness ranged from 19 to 26 mm in the apical segments, 11.0 mm in the basal left ventricular posterior wall, and 10.0 mm at the basal septum. No apical dyskinesis was visualized. The medial mitral annular velocity was attenuated at 8.0 cm/s (tissue Doppler). Two-dimensional strain analysis was later performed and revealed average segmental longitudinal strain of $-20$ and $-13\%$ in the basal and mid segments, respectively, and systolic lengthening (paradoxical strain) in the apical segments ($+16\%$, Figure 4).

Discussion

Prevalence of the apical variant of HCM in the Western world is low with a reported incidence of 1–2% of HCM. Traditionally, invasive procedures have been utilized for diagnosis with the confirmatory 'ace of spade' shaped cavity. Magnetic resonance tagged imaging has also been used to establish the diagnosis in patients with non-diagnostic echocardiograms.3 The utility of strain imaging in the apical variant of HCM has not been previously described.

In HCM, regional function is most abnormal in walls that are markedly hypertrophied, and this may represent a non-specific compensatory response to any mutation that impairs myofibrillar function. Tissue Doppler imaging (TDI) is a sensitive test to identify altered tissue velocities and reduced strain in affected segments of the LV. It can discriminate between HCM and athlete’s heart even before clear phenotypic changes develop.4

However, TDI strain is restricted to longitudinal strain measurements in the basal and mid segments (insonation angle dependency) or posterior wall assessment of radial strain. Segmental analysis of deformation and strain at the left ventricular apex is limited. With increasing insonation
angles as encountered in the apex, transverse thickening (positive strain) is measured instead of longitudinal shortening (negative strain). Two-dimensional strain or speckle tracking imaging, in contrast, tracks deformation between naturally occurring acoustic markers in the ultrasonic image in two dimensions, allowing one to study orthogonal components of strain independent of the insonation angle\(^1\); as a result, it is uniquely suited for the assessment of LV apical deformation. It is similar in concept to magnetic resonance imaging (MRI) tagging but unlike MRI, the persistence of tags in the imaging plane permits real-time calculation of frame-to-frame changes in myocardial motion, velocities, and derives deformation parameters such as strain.

We were able to rapidly and reproducibly demonstrate paradoxical strain in the apical segments (in the absence of apparent regional apical dyskinesis on conventional imaging) using 2D strain imaging, a finding that appears to support the diagnosis of apical HCM.

We also observed attenuated strain values in the mid-ventricular segments relative to the basal segments in contrast to the base-apex gradient in strain values reported in normal subjects using this technique.\(^1,2\) This finding appears to correlate with pathologic ventricular hypertrophy extending beyond the apex into the mid-ventricular segments, as apparent in the 2D apical images. Paradoxical strain (longitudinal systolic expansion) of the midseptum has been reported in patients with HCM using tissue Doppler strain imaging and directly relates to the degree of septal hypertrophy.\(^5\) In both our patients, paradoxical strain was observed in the most hypertrophied segments of the left ventricular apex.

The diagnosis of apical HCM can be challenging, necessitating additional expensive imaging modalities such as cardiac MRI, contrast-enhanced echocardiography, or left ventricular angiography. Two-dimensional strain, on the other hand, can be rapidly computed from conventional black and white echocardiographic images and appears to be a promising novel application, particularly suited for the diagnosis of suspected apical HCM. While clearly in need of further clinical validation, it could complement the role of contrast echocardiography and mitigate the need for more expensive or invasive imaging alternatives in patients with otherwise non-diagnostic or equivocal echo exams.

**References**