MR imaging in Hyperthrophic Cardiomyopathy: When CMR saves the day.

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Learning objectives

The purpose of our educational exhibit is to:

1. demonstrate the heterogeneity of hypertrophic cardiomyopathy (HCM) and illustrate the most common phenotypes of HCM;
2. familiarize the viewer with the role of advance cardiac imaging (in particular echocardiography and cardiac magnetic resonance imaging) in the evaluation of HCM;
3. emphasize the potential pitfalls when interpreting the images and the applications of cardiac magnetic resonance imaging in the evaluation of HCM.
Hypertrophic cardiomyopathy (HCM) is an autosomal dominant disease, clinically defined by the presence of left ventricular (LV) hypertrophy in the absence of any other detectable aetiology (e.g. hypertension or mild aortic stenosis with marked hypertrophy). The impact of advanced diagnostic imaging technologies in the diagnosis of HCM is great, particularly in the detection of certain phenotypes that would previously remain underdiagnosed.

Echocardiography is an excellent, easily accessible tool for the assessment of myocardial structure and function. However, there are certainly limitations. First of all it is an operator-dependent procedure. Secondly, the quality of imaging is dependent on the acoustic windows and not infrequently echocardiography is unable to depict the endocardial border, especially the anterolateral free wall of the left ventricle in the parasternal long-axis view and the apical short axis view. In addition suboptimal images limit accuracy and reproducibility of measurements (1).

There are several pitfalls in the detection of LV hypertrophy with echocardiography:

1. overdetection (false-positive): moderator band, oblique LV cut, left ventricular non-compaction syndrome;
2. underdetection (false-negative): poor endocardial definition of the LV apex, unusual pattern of left ventricular hypertrophy (anterolateral wall and apex).

Therefore; although echocardiography is the simplest and most readily available imaging modality for HCM screening, cardiac magnetic resonance imaging (CMRI) offers invaluable information for establishing the diagnosis particularly in cases when echocardiography results are inconclusive/ equivocal. CMRI is instrumental for the differential diagnosis of HCM as, amongst others, it is the most useful tool for tissue characterization. In selected patients with HCM, CMRI can be a useful tool in the risk stratification of patients with HCM.
Findings and procedure details

Phenotypes of HCM

HCM is actually more common than previously considered (1/500 individuals). It is particularly significant as it is one of the commonest causes of sudden cardiac death in young individuals, including young athletes. The morphologic expression of HCM is heterogeneous as HCM may affect any part of the left ventricle (2).

Asymmetric (Septal) HCM

This is the most common cause of the disease accounting approximately of 60-70% of HCM cases. In this phenotype the hypertrophy is asymmetric and typically involves the anteroseptum. The usual diagnostic criterion is a maximal LV wall thickness greater or equal to 15mm in the end-diastolic phase or a ratio of septal wall thickness to the thickness of the inferior wall greater than 1.5 at the mid-ventricular level (Fig. 1-5).

The presence of LVOT gradient with the patient at rest or during stress caused by systolic anterior motion of the mitral valve is important in order to distinguish between obstructive and non-obstructive forms of HCM. Peak LVOT gradients can be assessed by CMRI; however, assessment with echocardiography is currently a more accurate method, thanks to higher temporal resolution when measuring peak instantaneous velocities (Fig. 6-7).

Apical HCM

Apical HCM is diagnosed in the presence of one of the following (a) absolute wall thickness of >15mm or (b) ratio of apical to basal LV wall thickness 1.3-1.5. With CMRI the characteristic “spade-like” configuration of the LV cavity at end-systole (Fig. 12), which is caused by localized apical hypertrophy, can be appreciated on the vertical long-axis view. The role of echocardiography is important (Fig 8-10); however, the role of CMRI is undeniably particularly significant in apical HCM cases as the LV apex is frequently not properly visualized with echocardiography leading to false-negative results. Therefore; CMR is strongly recommended as the preferred imaging modality in apical HCM (Fig. 11-15). Fig. 14 demonstrates several crypts a finding which has been described in HCM and is clearly demonstrated in long axis CMRI views.

Concentric HCM
This type of HCM is characterized by concentric LV hypertrophy with a small LV cavity (Fig. 16). Differential diagnosis from other causes for LV hypertrophy may prove challenging without the use of CMRI. In particular the LGE imaging can be extremely useful in the characterization of the late gadolinium enhancement patterns of the hypertrophied myocardium.

**Mid ventricular HCM**

Midventricular HCM is characterized by hypertrophy of the LV wall at mid-level and is often associated with an apical aneurysm as a result of increased systolic pressures in the apex from mid-ventricular obstruction (Fig. 17-20). CMRI offers comprehensive views of the apical segments and allows the evaluation of the presence/absence of apical thrombus. The Fig. 17-20 are a characteristic example of a case of mid-ventricular 'burn-out' HCM.

**Differential diagnosis**

In cases of LV hypertrophy with insufficient explanation, accurate diagnosis of the underlying cause might prove difficult with echocardiography only used as an imaging modality, particularly in cases where HCM and hypertension co-exist. In these cases the significance of CMRI and tissue characterization is highlighted; approximately 80% of patients with HCM demonstrate hyperenhancement of the hypertrophied myocardial segments (3).

**Hypertensive Heart Disease**

This represents by far the greatest challenge in the differential diagnosis, particularly as there is considerable overlap between HCM and hypertensive heart disease. CMR allows accurate assessment of the LV systolic function which is in the normal range rather than hyperdynamic, the LV wall thickness which rarely exceeds 15mm. The LV myocardium rarely demonstrates hyperenhancement at late gadolinium enhancement imaging.

**Athlete’s Heart**

Athlete’s heart refers to morphologic changes in the heart, including increased LV mass, increased end diastolic cavity dimension and increased LV wall thickness. CMRI provides accurate assessment of various geometric indexes which aids the correct identification of athlete’s heart. Lack of areas of delayed hyperenhancement of the LV myocardium is another distinguishing factor. The differential diagnosis can prove particularly challenging in Afro-Caribbean athletes...
who frequently display a very abnormal ECG and in whom LVH is frequently more pronounced.

**Cardiac Amyloid**

In CMRI imaging, cardiac amyloidosis cases demonstrate a distinct pattern of late gadolinium hyperenhancement distributed subendocardially in a circumferential pattern. Moreover, involvement of all four chambers and the presence of pericardial and pleural effusions can be easily identified by CMR.

**Fabry’s Disease**

It is particularly difficult to distinguish Fabry's disease from HCM. History, clinical examination and involvement of other organ systems provide clues to the diagnosis. Some late gadolinium enhancement patterns have been described more frequently in Fabry's disease such as mid-wall LV fibrosis.
Fig. 1: Echocardiography: Parasternal long axis view in septal HCM.

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**Fig. 2:** CMR: Horizontal long axis cine imaging in a case of HCM with asymmetrical septal LV wall hypertrophy.

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**Fig. 3:** CMR: Vertical long axis cine imaging in a case of HCM with asymmetrical anterior and inferior LV wall hypertrophy.

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**Fig. 4:** CMR: Short axis cine imaging (left) in a case of septal HCM with marked hypertrophy of the anteroseptum. Following administration of gadolinium diffuse hyperenhancement of the hypertrophied segments is noted (right).

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**Fig. 5:** CMR: Horizontal long axis cine view of a case of septal HCM (right). Following administration of gadolinium mid-wall late enhancement of the basal septum is noted (left).

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**Fig. 6:** Echocardiography: 4-Chamber view in a case of HCM with systolic anterior motion of the mitral valve.

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**Fig. 7:** Doppler Echocardiography: 4-Chamber view in a case of HCM with systolic anterior motion of the mitral valve and mitral regurgitation.
Fig. 8: Echocardiography: 4-Chamber view in a case of apical HCM.
**Fig. 9:** Echocardiography: Parasternal long axis imaging in a case of apical HCM.

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Fig. 10: Echocardiography: Apical short axis view of a case with apical HCM. Obliteration of the apical LV cavity in end-systole.

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**Fig. 11:** CMR: Horizontal long axis cine imaging in a case of apical HCM.

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**Fig. 12:** CMR: Left ventricular outflow tract cine imaging in a case of apical HCM.

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Fig. 13: CMR: Short axis cine imaging of a case with apical HCM. Obliteration of the apical LV cavity in end-systole.

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Fig. 14: CMR: Left ventricular outflow tract cine imaging in a case of apical HCM with several crypts present.
**Fig. 15**: CMR: Horizontal long axis cine view of a case of apical HCM (left). Following administration of gadolinium diffuse hyperenhancement of the hypertrophied segments is noted (right).
**Fig. 16**: Echocardiography: 4-Chamber view of a case with concentric HCM.

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**Fig. 17:** CMR: Horizontal long axis cine imaging in a case of HCM with mid-ventricular LV wall hypertrophy and apical aneurysm.

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**Fig. 18:** CMR: Vertical long axis cine imaging in a case of HCM with mid-ventricular LV wall hypertrophy and apical aneurysm.
Fig. 19: CMR: Left ventricular outflow tract cine imaging in a case of HCM with mid-ventricular LV wall hypertrophy and apical aneurysm.

Fig. 20: CMR: vertical long axis cine imaging showing left ventricular hypertrophy in the mid-ventricular level and thinning and aneurysmal apical segments (left). Late gadolinium enhancement; vertical long axis view demonstrating patchy fibrosis of the hypertrophied segments and mid-wall fibrosis in the apical segments (right).
Conclusion

Establishing the diagnosis of HCM is crucial yet often challenging because of the technical limitations of transthoracic echocardiography while the heterogeneity of the disease makes assessment difficult. The role of CMR imaging is increasing and may eventually become the standard for the diagnosis of HCM, follow-up and risk stratification of HCM.

CMRI is particularly useful in the following settings (4):

1. Suspected HCM with equivocal echocardiographic findings.
2. HCM cases with apical or lateral LV wall hypertrophy and apical aneurysms.
3. Assessment of papillary muscles anatomy.
4. Differential diagnosis of other cardiomyopathies.
5. Emerging role in the preclinical detection of HCM and risk-stratification in established cases.
References


