Magnetic Resonance Imaging of Hypertrophic Cardiomyopathy

KH Tsang¹, WSW Chan², CK Shiu¹, JCY Lee¹, MK Chan¹

¹Department of Radiology and Imaging, Queen Elizabeth Hospital, Jordan, Hong Kong
²Department of Radiology and Organ Imaging, United Christian Hospital, Kwun Tong, Hong Kong

ABSTRACT

Hypertrophic cardiomyopathy is the most common genetically inherited cardiac disorder. It is characterised by a diffuse or segmental left ventricular hypertrophy. Its diagnosis is based largely on magnetic resonance imaging owing to its precise determination of myocardial anatomy for phenotype classification and risk stratification. This study discusses different phenotypes of hypertrophic cardiomyopathy on magnetic resonance imaging.

Key Words: Cardiomyopathy, hypertrophic; Heart; Magnetic resonance imaging

中文摘要

肥厚性心肌病的磁共振成像

曾劍鴻、陳施媛、蕭俊傑、李俊賢、陳文光

肥厚性心肌病（HCM）是最常見的基因遺傳心臟疾病。其特徵為擴散性或節段性左心室肥大，其診斷主要基於磁共振成像（MRI），因其能精確測定心肌解剖並作表型分類和風險分層。本文討論HCM在MRI上的不同表型。

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is the most common genetically inherited cardiac disorder with an autosomal dominant inheritance.¹,³ It is characterised by a diffuse or segmental hypertrophy of the left ventricle (LV), usually in the absence of other cardiac or systemic disease that leads to myocardial hypertrophy.¹ The main histological features are myocyte and myofibrillar disarray.¹,⁵ Diagnosis of HCM is based largely on magnetic resonance imaging (MRI), which can provide precise determination of the myocardial anatomy for phenotype classification and risk stratification (Figure 1). Delayed gadolinium-enhanced MRI of the myocardium shows tissue characterisation, specifically for identification of myocardial scar or fibrosis,¹,³ which is important in risk stratification. This study discusses different phenotypes of HCM on MRI.

PHENOTYPES

The incidence of HCM is about 0.2%.⁶ Clinical presentation varies from asymptomatic to dyspnoea, chest pain, syncope, or sudden cardiac death. The usual diagnostic criterion for HCM is an LV wall thickness of ≥15 mm measured in the end-diastolic phase (Figure 1).
Phenotypes are widely variable and heterogeneous; different segments of the LV can be affected.

Asymmetrical (septal) HCM is most common, accounting for 60% to 70% of all HCM. Hypertrophy of the anteroseptal LV myocardium is the most common pattern (Figure 3). The diagnosis is made when the septal wall thickness is $\geq 15$ mm or when the ratio of the septal wall thickness to the thickness of the inferior wall of the LV is $>1.5$ at the mid-ventricular level. Distinguishing the obstructive from non-obstructive forms is clinically important. The altered haemodynamic force along the LV outflow tract causes systolic anterior motion of the anterior mitral valve leaflet and the resulting leaflet-septal contact and obstructive physiology. Concomitant mitral regurgitation with the regurgitant jet directed posteriorly into the left atrium occurs secondary to incomplete leaflet apposition.

Apical HCM is characterised by myocardial hypertrophy predominantly affecting the LV apex. This results in a spade-like configuration of the LV cavity on vertical long-axis view at the end-diastolic phase (Figure 4). The diagnosis is made when the apical wall thickness is $>15$ mm or the ratio of apical to basal LV wall thickness is 1.3 to 1.5. This variant is seen more frequently in Japanese than in Western populations (about 25% vs. 2% of all HCM). It is frequently complicated by hypertension but rarely associated with sudden cardiac death.

Concentric (symmetrical) HCM accounts for 42% of all HCM and is characterised by concentric or symmetrical LV hypertrophy in the absence of...
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Secondary causes (Figure 5). The myocardial thickness is >15 mm and is less commonly seen in other causes that result in diffuse hypertrophy such as hypertensive disease. The LV cavity dimension is reduced in a concentric manner.

Mid-ventricular HCM is a rare variant of asymmetric HCM and is characterised by hypertrophy of the middle third of the LV wall (Figure 6), with mid-LV cavity narrowing with a dumbbell configuration. In severe cases, it is associated with an apical dilatation.
secondary to increased systolic pressure in the apex as a result of mid-ventricular obstruction. In about 10% of patients, there is progression to a burned-out apex secondary to ischaemia with apical aneurysm formation and late myocardial enhancement after gadolinium administration. Mass-like HCM is due to the focal segmental location
of the myocardial disarray and fibrosis (Figure 7). It is differentiated from a genuine myocardial mass by its homogeneous signal intensity and perfusion signal parallel to the adjacent normal myocardium. The myocardial tagging technique shows contractility in the hypertrophied segment, which is absent in myocardial neoplasm.

DISCUSSION
Myocardial fibrosis or scarring can be detected by cardiac MRI in 33% to 86% of patients with HCM. As opposed to subendocardial distribution of late gadolinium enhancement in myocardial infarcts, the pattern of late gadolinium enhancement in HCM usually appears as small punctate, patchy hyperenhancement with a mid-wall distribution (Figure 8). The regional location of late gadolinium enhancement within the hypertrophied segment is suggestive of underlying HCM.

Risk factors that can be detected by cardiac MRI include myocardial wall thickness ≥30 mm, presence of LV outflow tract obstruction or systolic anterior motion, presence of wall fibrosis, myocardial wall perfusion...
defect, LV dilatation, and decreased ejection fraction.

Between January 2010 and May 2015 at Queen Elizabeth Hospital in Hong Kong, 29 men and 15 women aged 36 to 85 (mean, 63) years were diagnosed by MRI with septal HCM (n = 31), apical HCM (n = 6), symmetrical HCM (n = 5), mid-ventricular HCM (n = 1), and mass-like HCM (n = 1). The mean hypertrophied LV wall thickness was 20.1 (standard deviation, 4.2; range, 15-36) mm. Most patients had mid-wall fibrosis, probably owing to old age. Systolic anterior motion of the anterior mitral valve leaflet was identified in 10 of the 31 patients with septal HCM. Late mid-myocardial gadolinium enhancement was identified in 41 patients.

CONCLUSION
Cardiac MRI is useful for diagnosing and risk stratification of HCM. Familiarisation with its MRI appearance can help make a more accurate diagnosis.

REFERENCES