

THE MAGNETIC RESONANCE IMAGING CHARACTERISTICS OF HYPERTROPHIC CARDIOMYOPATHY

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SUMMARY

Objectives: Describe the magnetic resonance imaging characteristics of hypertrophic cardiomyopathy.

Subjects and methods: A prospective, cross-sectional study on 30 patients with hypertrophic cardiomyopathy at the Vietnam Heart Institute, undergoing cardiac magnetic resonance imaging at the Center for Radiology, Bach Mai Hospital from July 2021 to September 2022.

Results: Maximum wall thickness is 37 mm, mid anteroseptal: 21.8 ± 6.17 mm; mid anterolateral: 20.6 ± 3.98 mm, basal inferolateral: 15.9 ± 1.77 mm, basal inferior: 15.9 ± 0.71 mm. 6.7% hypertrophy of both ventricles; 13.3% concentric hypertrophy; 40% diffuse eccentricity; 30% deviation of the entire interventricular septum and 3.3% of the apex. Mean ejection fraction: $65.58 \pm 9.02\%$; mean left ventricular mass: 181.98 ± 55.14 g, mean left ventricular outflow tract diameter: 9.56 ± 4.69 mm and the ratio of left ventricular outflow tract diameter to annulus diameter host: 0.46 ± 0.23 mm. 46.7% presented late gadolinium enhancement and 46.7% having signs of SAM.

Conclusion: The phenotypic characteristics, wall thickness, left ventricular mass, ejection fraction, late gadolinium enhancement and some other abnormalities associated with magnetic resonance imaging provide useful data for the treatment of hypertrophic cardiomyopathy.

Keywords: *hypertrophic cardiomyopathy, magnetic resonance imaging characteristics*

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I. INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is still considered a burden of disease worldwide, with an estimated prevalence of 0.2% of the population (1/500), being one of the most common causes of sudden cardiac death, especially in young patients under 35 years old [1]. Therefore, early and accurate diagnosis of HCM has a decisive significance in the direction of care, timely treatment, and prevention of complications. Clinical examination and initial laboratory tests upon admission are not always definitive of the diagnosis, especially in patients whose ultrasound window is limited, signs of hypertrophy are unclear, and it is easy to miss the diagnosis [2]. However, in recent years, cardiac magnetic resonance imaging (MRI) has emerged as an imaging modality that overcoming the limitations of echocardiography along with many apparent advantages such as the ability to visualize myocardium and high reproducibility. MRI is considered the gold standard for quantifying left ventricular volume, mass, wall thickness, left ventricular function, and phenotypic classification. MRI has been recommended to aid echocardiography in definitive diagnosis, differential diagnosis, and treatment. However, in Vietnam, so far, there are few studies on HCM. Therefore, we carried out the study to characterize the magnetic resonance imaging characteristics of HCM, thereby contributing to the diagnosis and prognosis and giving an appropriate treatment strategy.

II. SUBJECT AND METHODS

2.1. Subject

All patients diagnosed with HCM at the Vietnam National Heart Institute underwent cardiac MRI at the Radiology Center, Bach Mai Hospital, from July 2021 to September 2022.

Selection criteria

- Patients can be clinically suspected or diagnosed with HCM according to the European Society of Cardiology 2014 guidelines.
- The patient consented to participate in the study.

Exclusion criteria

- Patients with MRI contraindications include metal foreign bodies (in the orbit, skull, heart ...) and implantable medical devices (hearing aid, pacemaker...).
- Patients had medical, surgical, metabolic, or occupational conditions that can be responsible for myocardial hypertrophy, such as hypertension and amyloidosis
- Patients were allergic to contrast agents or had severe renal failure.
- Magnetic resonance images had non-diagnostic quality.
- Patients had claustrophobia or could not cooperate during MRI scans.

2.2. Methods

2.2.1. Research design: a descriptive cross-sectional study

2.2.2. Sampling method and sample size: convenience sampling method. The study sample consisted of 30 patients

2.2.3. Research equipment

- SIGNA Architect 3.0T MRI scanner (GE Healthcare, USA).
- Images were processed on an MR Workspace (Phillips, Netherlands) and CVI42 software (Circle Cardiovascular Imaging, Canada).
- MRI contrast agent: DOTAREM (Guerbet, France).

2.3. MRI protocols

- Multiple plane localizers, including axial, coronal, sagittal, 2-chamber, 3-chamber, 4-chamber, short axis, and left ventricular outflow tract (LVOT) views were taken initially.
- Taking 2-chamber, 4-chamber, short-axis cine sequences (8-10 slices) from the base to the apex to evaluate left ventricular mass, EF, and left ventricular end-systolic and end-diastolic volume, wall thickness, papillary muscle abnormalities. 3-chamber cine and LVOT sequences to assess SAM (Systolic anterior motion), mitral leaflets length, and LVOT diameter.
- Then injecting Dotarem at 0.2 mmol/kg intravenously, 2-3 ml/sec, followed by 20-25 ml saline. The delayed

phase was acquired with a delay of 10 minutes of initiation of the contrast injection. Determining optimal TI (time inversion) with deletion of standard myocardial signal, using reversible recovery (IR) pulse sequence with TI time, take late gadolinium enhancement images with one slice on each 2-chamber, 4-chamber views and eight slices on short-axis view covering from base to apex.

2.4. Data collection and Statistical analysis

- Collected data according to the research data collection form.

- Data were processed using IBM SPSS Statistics for Windows, version 20.0 (IBM Corp., Armonk, N.Y., USA).

2.5. Ethics in research

Hanoi Medical University Institutional Ethical Review Board has approved the research. All patients thoroughly explained the purpose of the research and could withdraw from the study at any time based on their wishes. All patients' personal information was kept confidential and used for research only.

III. RESULTS

The study consisted of 30 patients, including 18 males (60%). The mean age of the patients was 51.7 ± 19.56 years old, with an age range of 19–84 years. The mean BMI was 22.6 ± 3.44 kg/m²; the mean heart rate was 79.1 ± 23.98 beats per minute.

Table 1. Clinical characteristics of the study sample (n=30)

Clinical characteristic	n	Percentage (%)
Family history of HCM	6	20
Family history of sudden cardiac death	4	13.3
Family history of implantable cardioverter-defibrillator	1	3.3
Syncope/ presyncope/fainting	7	23.3
Dyspnea	23	76.7
Chest pain	23	76.7

20% of patients had a family history of HCM, 13.3% of

patients had a family history of sudden death, and 3.3% of patients had a family history of implantable cardioverter-defibrillator. Most patients had symptoms of dyspnea (76.7%), chest pain (76.7%), syncope/transient syncope/fainting (23.3%).

Table 2. The LV wall thickness of hypertrophic segments according to the American Heart Association 17-segmented model

Segment	Min (mm)	Max (mm)	Mean (mm)
Maximum wall thickness	15.3	37.0	22.9 ± 5.6
Basal anterior (n=15)	15.1	30.4	20.1 ± 3.9
Basal anteroseptal (n=22)	1.3	24.2	18.9 ± 2.49
basal inferoseptal (n=14)	13.8	23.2	17.1 ± 3.05
basal inferior (n=5)	13.3	18.0	15.9 ± 1.77
basal inferolateral (n=5)	15	16.8	15.9 ± 0.71
basal anterolateral (n=10)	14	18.6	16.2 ± 1.28
mid anterior (n=14)	14.4	23.0	18.2 ± 2.28
mid anteroseptal (n=25)	14	37.0	21.8 ± 6.17
mid inferoseptal (n=24)	15.1	29.3	20.6 ± 3.98
mid inferior (n=14)	14.5	20.4	17.1 ± 2.03
mid inferolateral (n=4)	15.4	20.6	17.6 ± 2.39
mid anterolateral (n=10)	15.0	18.6	16.5 ± 1.1
apical anterior(n=8)	15	27.6	18.5 ± 4.17
apical septal (n=9)	15.0	21.2	17.2 ± 2.33
apical inferior (n=9)	14.8	21.7	16.9 ± 2.09
apical lateral (n=10)	15.0	21.6	17.7 ± 2.63

The maximal wall thickness was 37mm, and the mid-anteroseptal segment had the most significant mean thickness of 21.8 ± 6.17 mm, followed by the mid-inferoseptal segment (20.6 ± 3.98 mm). The segments with the minimal thickness were the basal inferior segment (15.9 ± 1.77 mm) and the basal inferolateral segment (15.9 ± 0.71 mm).

Table 3. HCM phenotypes on cardiac MRI (n=30)

Phenotype			Total	
			n	Percentage
Left ventricle (n=28)	asymmetric HCM (n=24)	diffuse asymmetric HCM	12	40.0
		septal HCM	9	30.0
		mid-septal HCM	2	6.7
		Apical HCM	1	3.3
	concentric HCM		4	13.3
Both left and right ventricles			2	6.7

Of the 30 patients, 6.7% had both right and left ventricular hypertrophy; 13.3% had concentric HCM. Diffuse asymmetric HCM accounted for the highest rate (40.0%),

followed by septal HCM (30.0%). 6.7% had midventricular HCM, and only one patient (3.3%) had apical HCM.

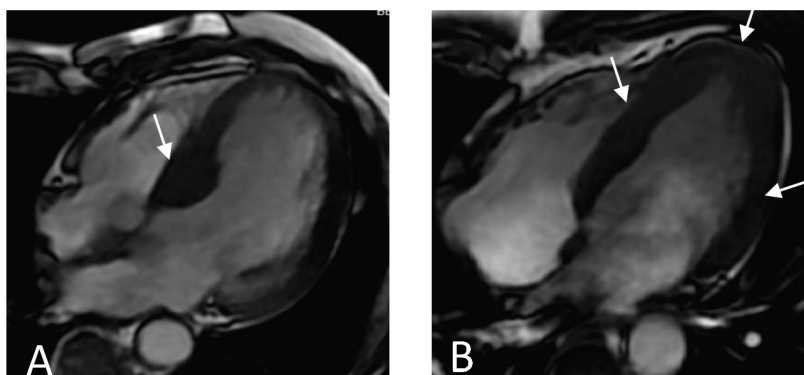


Figure 1. Cine 4-chamber view in end-diastolic phase.
(A) A 70-year-old male had mid-septal HCM (arrows).
(B) A 46-year-old male had concentric HCM (arrows)

Table 4. MRI characteristics of the study sample

MRI characteristic	Min	Max	$\bar{X} \pm SD$
Left ventricular (LV) mass (g)	83.17	303.91	181.98 $\pm$ 55.14
EF (%)			
LVEF (%)	45.4	78.67	65.58 $\pm$ 9.02
VLAi (ml/m ²)	19.12	144.9	51.89 $\pm$ 27.46
Left ventricular outflow tract (LVOT) diameter (mm)	3	18.2	9.56 $\pm$ 4.69
Aortic annulus diameter (mm)	16.1	32.0	21.08 $\pm$ 3.23
LVOT/Ao diameter ratio	0.14	0.83	0.46 $\pm$ 0.23

The mean left ventricular mass was 181.98 $\pm$ 55.14g, the mean EF was 65.58 $\pm$ 9.02%, mean VLAi was 51.89 $\pm$ 27.46 ml/m². The mean LVOT and aortic annulus

diameter were 9.56 $\pm$ 4.69 mm and 21.08 $\pm$ 3.23 mm, respectively. The lowest LV/Ao ratio was 0.14, the highest was 0.83, and the mean was 0.46 $\pm$ 0.23.

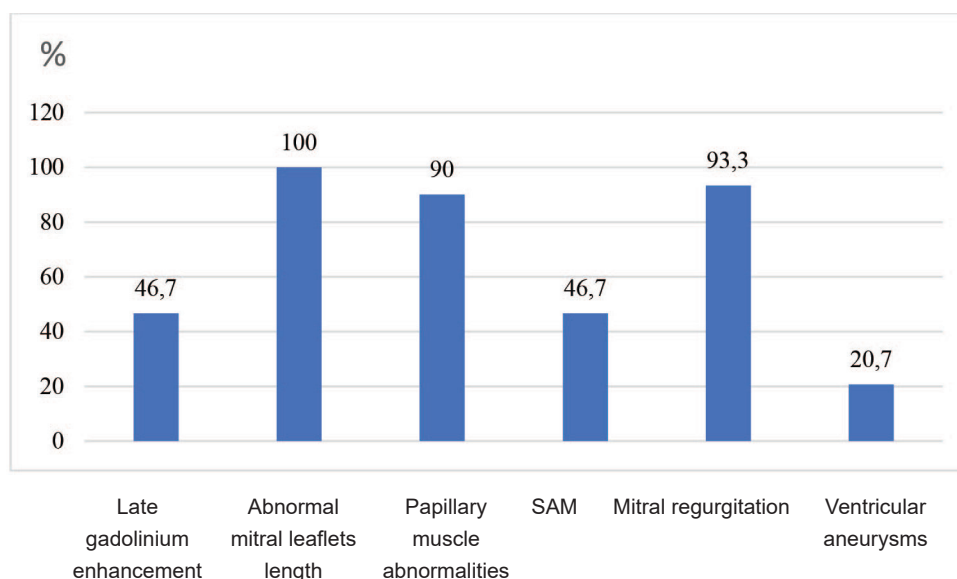


Figure 1. Other MRI findings.

About half of the patients had late gadolinium enhancement (46.7%) and SAM (46.7%). All patients had abnormal mitral leaflets length, 90% had papillary muscle abnormalities, 93.3% had mitral regurgitation, and 20.7% had ventricular aneurysms.

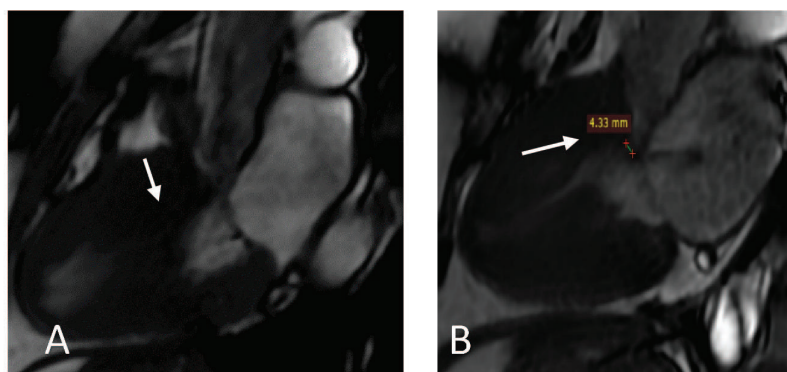


Figure 2. A 72-year-old female had severe LOVT obstruction due to SAM on cine 3-chamber view (A) and LVOT view (B).

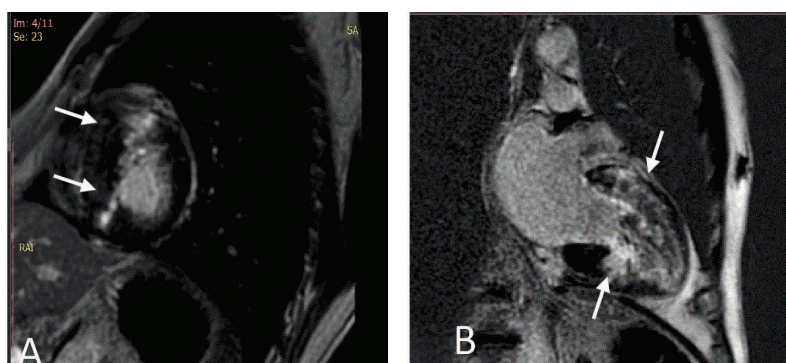


Figure 3. A 55-year-old male had late gadolinium enhancement with transmural pattern on short-axis view (arrows in A) and mid-wall pattern on 2-chamber view (arrows in B).

IV. DISCUSSION

Our study consisted of 30 HCM patients with an average age of 51.7 ± 19.56 years old, male/female ratio was 1.5, similar to Corona et al [3]. HCM is caused by an autosomal dominant mutation, which can be transmitted to the next generation. People have a 50% risk of carrying a mutated gene if their father and/or mother have HCM [4]. In our study, 20% of the patients had a family history of HCM, 13.3% had a family history of sudden cardiac death, and 3.3% had a family history of implantable cardioverter-defibrillator. This result was higher than the study of Wenxiu Chan et al [5], with these rates being 13.3%, 3.9%, and 2.2%, respectively, moreover higher than the results of Alaa Alashi et al.6 found that patients with a family history of HCM and sudden cardiac death accounted for 2% and 5%, respectively. We assumed that this difference was due to our small sample size, different study subjects, and locations of research.

Syncope, dyspnea, and chest pain were the most common symptoms in our study. Most patients had dyspnea and chest pain, and about 1/4 of the patients had syncope. Our results were consistent with other studies. Compared with Alashi et al [6], our study was similar in dyspnea, almost NYHA class II, but their chest pain and syncope rates were lower than our results (19% and 12%, respectively). It was due to different ages, and our study subjects were mainly patients with heart failure.

Cardiac MRI helps to assess the myocardial hypertrophy of each segment easily. In our study, almost patients had left ventricular hypertrophy; only two patients had both ventricular hypertrophy. Concentric HCM was less common than asymmetric HCM. The most common asymmetric phenotype was diffuse hypertrophy (40%), septal hypertrophy (30%), and mid-septal hypertrophy (6.7%), and only one patient had apical HCM. The phenotype in our study was consistent with the study of Mai Thanh Thao et al [7], and the epidemiology of HCM mainly in the left ventricle [9].

Many studies compared the measurement of wall thickness between ultrasound and magnetic resonance [10]. Toshimitsu Tsugun et al. measured wall thickness in 56 HCM patients and showed that the mean posterior

wall and maximum wall thickness were 11.5 mm and 20.1mm, respectively [11]. These results were lower than our study due to the difference in subjects and stages. Accurate assessment of wall thickness helps stratify and identify patients at high risk for arrhythmias which may need an implantable cardioverter-defibrillator. Therefore, wall thickness measurement should be a fundamental and essential part of the initial evaluation for all HCM patients.

The effectiveness of the treatment depends on many factors, in which the stage of disease plays an important role. The reduced LV ejection fraction indicates that the patient is already in the advanced stage and at risk of developing decompensated heart failure. Our results (Table 4) were similar to Toshimitsu Tsugun et al [8], Mai Thanh Thao et al [7]. In our study, the mean LV ejection fraction approached the upper limit of systolic dysfunction, so MRI and clinical monitoring were necessary during treatment. The mean LV mass was higher than normal because the patients were predominantly late-stage. Accurate assessment of myocardial mass could help clinicians better stratify risk. In addition, evaluating the obstruction degree also plays an essential role in the treatment and prognosis. Jens Vogel-Claussen et al.9 studied 92 HCM patients and assessed the degree of obstruction based on the left ventricular outflow tract/aortic valve diameter ratio showed the average was 0.41, the ratio was 0.6 ± 0.13 in the non-obstructive HCM group which was higher than latent obstructive HCM (0.41 ± 0.16) and obstructive HCM (0.24 ± 0.09). This result was lower than our result due to different study populations.

In addition to the ventricular indices, the left atrial volume index plays a role in assessing diastolic function - a predictor of atrial fibrillation and stroke. MRI can accurately evaluate the left atrial volume index. In a study by Sainan Cheng et al [10] on 63 end-stage HCM patients, the left atrial volume index was 96.9 ± 51.0 ml/m², much higher than our results because some patients in our study had early stages.

Papillary muscle abnormalities and elongated mitral leaflets are essential factors contributing significantly to the obstructive condition in HCM. Our study found that most patients had mitral valve and papillary muscle

abnormalities, and nearly half had SAM. SAM is the most characteristic movement anomaly of HCM, with a specificity of up to 98%, resulting in LVOT obstruction. Our results are lower than Dang Thi Linh et al. (88.8%) but still within the limits of previous studies (31-61%) [11]. The difference was due to our study using MRI while Dang Thi Linh used ultrasound, along with different research subjects and times. Based on the high resolution of the MRI, our study detected that 17.8% of patients had left ventricular aneurysms that were not seen on previous ultrasounds due to limited observation and ultrasound window.

An outstanding advantage of MRI over echocardiography is the detection of myocardial fibrosis seen as late gadolinium enhancement, which is considered a

characteristic of HCM. The degree of late gadolinium enhancement has a significant role in prognosis, predicting progression, and risk factors for ventricular arrhythmias, ventricular dilatation, and sudden cardiac death. In our study, nearly half of the patients had late gadolinium enhancement, lower than Mai Thanh Thao et al. (88.9%) due to the difference in study sample size, location, time, and subjects at different disease stages [7].

V. CONCLUSION

Cardiac MRI is necessary for diagnosis and prognosis. Assessment of phenotype, wall thickness, left ventricular mass, LV ejection fraction, late gadolinium enhancement, and some other abnormalities on MRI provide valuable data for the treatment of HCM accurately and efficiently.

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Received: 11/10/2022 . Assessed: 11/10/2022. Reviewed: 19/10/2022. Accepted: 10/12/2022