Rev Port Cardiol. 2014;33(5):261–267

Revista Portuguesa de
Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

ORIGINAL ARTICLE

Phenotypic expression in hypertrophic cardiomyopathy and late gadolinium enhancement on cardiac magnetic resonance☆

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Received 9 June 2013; accepted 19 October 2013
Available online 18 June 2014

KEYWORDS
Hypertrophic cardiomyopathy;
Late gadolinium enhancement;
Cardiac magnetic resonance;
Prognosis

Abstract
Introduction and Aim: The prognostic value of late gadolinium enhancement (LGE) for risk stratification of hypertrophic cardiomyopathy (HCM) patients is the subject of disagreement. We set out to examine the association between clinical and morphological variables, risk factors for sudden cardiac death and LGE in HCM patients.
Methods: From a population of 78 patients with HCM, we studied 53 who underwent cardiac magnetic resonance. They were divided into two groups according to the presence or absence of LGE. Ventricular arrhythmias and morbidity and mortality during follow-up were analyzed.
Results: Patients with LGE were younger at the time of diagnosis (p=0.046) and more often had a family history of sudden death (p=0.008) and known coronary artery disease (p=0.086). On echocardiography they had greater maximum wall thickness (p=0.007) and left atrial area (p=0.037) and volume (p=0.035), and more often presented a restrictive pattern of diastolic dysfunction (p=0.011) with a higher E/E′ ratio (p=0.003) and left ventricular systolic dysfunction (p=0.038). Cardiac magnetic resonance supported the association between LGE and previous echocardiographic findings: greater left atrial area (p=0.029) and maximum wall thickness (p<0.001) and lower left ventricular ejection fraction (p=0.056). Patients with LGE more often had an implantable cardioverter-defibrillator (ICD) (p=0.015). At follow-up, no differences were found in the frequency of ventricular arrhythmias, appropriate ICD therapies or mortality.
Conclusions: The presence of LGE emerges as a risk marker, associated with the classical predictors of sudden cardiac death in this population. However, larger studies are required to confirm its independent association with clinical events.

☆ Please cite this article as: Caetano F, Botelho A, Trigo J, et al. Expressão fenotípica da miocardiopatia hipertrófica e realce tardio na ressonância magnética cardíaca. Rev Port Cardiol. 2014;33:261–266.

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PALAVRAS-CHAVE
Miocardiopatia hipertrófica;
Realce tardio;
Ressonância magnética cardíaca;
Prognóstico

Expressão fenotípica da miocardiopatia hipertrófica e realce tardio na ressonância magnética cardíaca

Resumo
Introdução e objetivos: O valor prognóstico do realce tardio na estratificação dos doentes com miocardiopatia hipertrófica é controverso. Este trabalho pretende avaliar a associação entre a presença de realce tardio na ressonância magnética cardíaca e características clínicas, imagiológicas e prognósticas em doentes com miocardiopatia hipertrófica.
Métodos: De 78 doentes com miocardiopatia hipertrófica avaliámos retrospetivamente 53, que realizaram ressonância cardíaca. Os doentes foram divididos em dois grupos, conforme a presença ou ausência de realce tardio. Foi feito seguimento clínico referente a disritmia ventricular e a morbi-mortalidade.
Resultados: Os doentes com realce tardio eram mais jovens à data do diagnóstico (p=0,046), mais frequentemente tinham antecedentes familiares de morte súbita (p=0,008) e de doença coronária (p=0,086). No ecocardiograma apresentavam maior espessura parietal máxima (p=0,007); área (p=0,037) e volume indexado da aurícula esquerda (p=0,035); maior frequência de padrão restritivo de disfunção diastólica (p=0,011), com relação E/E' mais elevada (p=0,003); e disfunção sistólica do ventrículo esquerdo (p=0,038). A ressonância validou as alterações ecocardiográficas associadas à presença de realce tardio: maior área da aurícula esquerda (p=0,029); espessura parietal máxima (p<0,001) e menor fração de ejeção do ventrículo esquerdo (p=0,056). Os doentes com realce tardio mais frequentemente eram portadores de CDI (p=0,015); não havendo diferenças na frequência de episódios de disritmia ventricular, terapias apropriadas de CDI ou mortalidade no seguimento clínico.
Conclusões: A presença de realce tardio surge como um marcador de risco, associando-se a fatores já reconhecidos como preditores de morte súbita nesta população. A sua associação independente a eventos clínicos exige o estudo de populações de maior dimensão.

List of abbreviations

AVB	atrioventricular block
CCBs	calcium channel blockers
CMR	cardiac magnetic resonance
HCM	hypertrophic cardiomyopathy
HR	hazard ratio
ICD	implantable cardioverter-defibrillator
LA	left atrial
LGE	late gadolinium enhancement
LV	left ventricular
LVEF	left ventricular ejection fraction
LVH	left ventricular hypertrophy
OR	odds ratio
NYHA	New York Heart Association

Introduction

Hypertrophic cardiomyopathy (HCM) is a relatively common hereditary disease with a prevalence of 1/500 population, characterized by complexity and a wide spectrum of phenotypic expression and natural history.[1,2]

Sudden death is the leading cause of mortality in young HCM patients, although the risk is low (about 1%/year); an implantable cardioverter-defibrillator (ICD) is the only effective preventive measure.[3] Risk stratification algorithms for predicting sudden death in this population have low positive predictive value and are unclear in their definition of risk factors.[3,4]

Cardiac magnetic resonance (CMR) has become an essential exam in the morphological and functional assessment of HCM. However, the prognostic value of late gadolinium enhancement (LGE) in identifying potentially arrhythmogenic areas of endomyocardial fibrosis is the subject of disagreement.[5]

The aim of this study was to examine the association between LGE on CMR and clinical, imaging and prognostic characteristics in a Portuguese population of HCM patients.

Methods

Study population

We performed a retrospective analysis of HCM patients followed regularly as outpatients in the cardiology department of a central hospital who underwent CMR with LGE study.

Of a population of 78 patients with HCM, 61 underwent CMR. The reasons for not undergoing CMR were the usual ones of ICD or pacemaker, claustrophobia, or patient refusal.

The definition of HCM was based on the classic echocardiographic criteria (left ventricular hypertrophy [LVH] without dilatation, maximum wall thickness $\geq$15 mm) and exclusion of other systemic or local causes of hypertrophy.