

**The Role Of Delayed Enhancement Cardiac
Magnetic Resonance Imaging Versus Two
Dimensional Echocardiography In The
Evaluation Of Hypertrophic
Cardiomyopathy Disease In Pediatrics**
Thesis

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MD degree in Radiology*

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Abstract

In this study including ischemia, microvascular dysfunction, and obstruction. However, recent observations have challenged this interpretation showing that the stimulus for myocardial fibrosis is an early manifestation of sarcomere-gene mutations that may be present before occurrence of cardiac morphological features of HCM. Myocardial fibrosis is present in the majority of patients with overt HCM, and corresponds with impairment in myocardial energy metabolism, and correlates with the severity of ventricular dysfunction. Use of late Gd imaging has become a well-accepted technique to depict focal myocardial fibrosis correlating well with the autopsy findings. There is an increasing body of evidence that the presence and extent of myocardial fibrosis, in patients with HCM, is an independent predictor of adverse outcome, i.e., SCD, sustained ventricular tachycardia or fibrillation, and heart failure. Also in patients with no, or minimal symptoms, late Gd imaging adds prognostic value to conventional criteria for risk stratification. Moreover, myocardial fibrosis is related not only to an increased risk for SCD, but also to the presence of progressive disease and prediction of systolic and diastolic dysfunction

Key word

CMR-MRI-DCM-SCD-HCM

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Table of Contents

Introduction.....	1
Objectives & Aim of Study.....	3
Pediatric Hypertrophic Cardiomyopathy.....	4
Physics.....	49
Cardiac Magnetic Resonance (CMR).....	56
Patients and Methods.....	65
Results.....	71
Representative cases.....	85
Discussion.....	95
Study Limitations.....	103
Conclusion.....	104
Recommendations.....	105
References.....	106
Summary.....	121

List of figures & tables

Figures

Figure 1. Myocardial fiber disarray in hypertrophic cardiomyopathy. (Sherrid MV. *Pathophysiology and Treatment of Hypertrophic Cardiomyopathy. Progress in Cardiovascular Diseases.* 2006; 49: 123–151)

Figure 2. Histopathology from surgical specimens of 3 patients with obstructive HCM. (Sherrid MV. *Pathophysiology and Treatment of Hypertrophic Cardiomyopathy. Progress in Cardiovascular Diseases.* 2006; 49: 123–151)

Figure 3. Comparison of survival free from HCM-related cardiac death, in patients with obstructive and nonobstructive HCM. (Maron MS, Olivotto I, Betocchi S, Casey SA, Lesser JR, Losi MA, Cecchi F, Maron BJ. *Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophy cardiomyopathy. N Engl J Med.* 2003b; 348:295–303.)

Figure 4. Systolic anterior motion of the mitral valve, drawn from apical 5 chamber view, as it proceeds in early systole. (Sherrid MV, Chu CK, Delia E, Mogtader A, Dwyer EM Jr. *An echocardiographic study of the fluid mechanics of obstruction in hypertrophic cardiomyopathy. J Am Coll Cardiol.* 1993 Sep;22(3):816-25.)

Figure 5. Hypertrophic cardiomyopathy. (Image courtesy of Michael E. Zevitz, MD)

Figure 6. ECG of a 16-year-old with hypertrophic cardiomyopathy (HCM) (<http://emedicine.medscape.com/article/890068-overview>)

Figure 7. Dark-blood versus bright-blood imaging in a patient with LV non-compaction cardiomyopathy (LVNC). (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI.* Springer. 2012)

Figure 8. Focal form of hypertrophic cardiomyopathy (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI.* Springer. 2012)

Figure 9. Asymmetrical septal HCM. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI.* Springer. 2012)

Figure 10. Extreme form of hypertrophic, obstructive cardiomyopathy in a young male teenager. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 11. Apical form of HCM (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 12. Spiral nature of HCM. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 13. Comprehensive MRI in severe form of obstructive hypertrophic cardiomyopathy. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 14. Deep muscular clefts in inferoseptal LV wall (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 15. Asymmetrical septal HCM with LVOT obstruction. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 16. Assessment of hemodynamic compromise in LVOT by PC-MRI (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 17. “Venturi”-effect in hypertrophic (obstructive) cardiomyopathy. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 18. Typical enhancement pattern in HCM. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 19. Relation between myocardial deformation and myocardial scarring in HCM. (Bogaert J, Taylor AM, Van Kerckhove F, Dymarkowski S. *Use of inversion-recovery contrast-enhanced MRI technique for cardiac imaging: spectrum of diseases*. Am J Roentgenol AJR. 2004; 182:609–615).

Figure 20. Myocardial enhancement in apical HCM. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 21. Spontaneous evolution of asymmetric septal HCM (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 22. Asymmetric septal obstructive HCM. (Bogaert J, Taylor AM, Van Kerckhove F, Dymarkowski S. *Use of inversion-recovery contrast-enhanced MRI technique for cardiac imaging: spectrum of diseases*. *Am J Roentgenol* AJR. 2004; 182:609–615).

Figure 23. Inversion recovery curve (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 24. Late Gd image of an inferior myocardial infarct. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 25. Phase-sensitive image reconstruction (PSIR) technique (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 26. Impact of inversion time on late Gd image quality. (Bogaert J, Dymarkowski S, Taylor AM, Muthurangu V. *Diagnostic Imaging: Clinical Cardiac MRI*. Springer. 2012)

Figure 27. MRI Knee coils (A), Cardiac coil (B) (http://www.healthcare.Philips.Com/main/products/mri/options_upgrades/coils)

Figure 28. LV: left ventricle, SA: Sagittal, 2C: 2 chamber, RVOT: right ventricular out flow tract. (Fratz S, Chung T, Greil GF, Samyn MM, Taylor AM, Buechel VER, Yoo SJ, Powell AJ. *Guidelines and protocols for cardiovascular magnetic resonance in children and adults with congenital heartdisease: SCMR expert consensus group on congenital heart disease*. *J Cardiovasc Magn Reson*. 2013; 13;15:51)

Figure 29. Retrospective ECG Gating (www.med-ed.virginia.edu/courses/rad/cardiacmr/Techniques/Cine).

Figure 30. Dynamic SSFP Short-axis Images (*Weber OM, Higgins CB. MR Evaluation of Cardiovascular Physiology in Congenital Heart Disease: Flow and Function. Journal of Cardiovascular Magnetic Resonance. 2006; 8:607–617*)

Figure 31. Endsystolic Phase Contour Tracing (*Buechel EV, Kaisel T, Jackson C, Schmitz A, Kellenberger CJ. Normal right- and left ventricular volumes and myocardial mass in children measured by steady state free precession cardiovascular magnetic resonance. J Cardiovasc Magn Reson. 2009; 1:11-19*)

Figure 32. Screen-shot showing quantification of percentage of delayed hyperenhancement done using Diagnosoft software.

Figure 33. Gender distribution among study group

Figure 34. Family history of HCM among study group

Figure 35. NYHA class distribution among study group

Figure 36. NYHA class distribution among study group correlated with severity of pressure gradient across the left ventricular outflow tract (PG LVOT)

Figure 37. NYHA class distribution among study group correlated with the percentage of delayed enhancement (DE)

Figure 38. Delayed enhancement distribution among study group

Figure 39. Comparisons between fibrosis & no-fibrosis groups were done regarding New York Heart Association Functional Classification (NYHA) & severity of pressure gradient across the left ventricular outflow tract (PG LVOT).

Figure 40. Distribution of pressure gradient across the left ventricular outflow tract (PG LVOT) among study group

Figure 41. Unpaired *t* tests comparison between obstructive and non-obstructive groups regarding the severity of NYHA class.

Figure 42. Fibrosis detected among study group by echo.

Figure 43. Pearson correlation between delayed enhancement percentage (DE%) & severity of pressure gradient across the left ventricular outflow tract (PG LVOT)

Fig. 44 Asymmetric septal hypertrophy: axial cine image.

Fig. 45 End-systole & end-diastole short axis cine image showing almost total obliteration of the ventricular lumen.

Fig. 46 Quantification of percentage of delayed enhancement.

Fig. 47 Asymmetric biventricular hypertrophy.

Fig. 48 Inflow-outflow & LVOT cine images showing SAM & subaortic systolic jet.

Fig. 49 Quantification of percentage of delayed enhancement.

Fig. 50 Mid cavity systolic jet denoting mid cavity obstruction.

Fig. 51 Short axis IR post contrast delayed images showing patchy mid-wall faint enhancement.

Fig. 52 Quantification of percentage of delayed enhancement.

Fig. 53 Axial cine image showing asymmetric septal hypertrophy, SAM & mitral regurge.

Fig. 54 Focal mid-wall enhancement.

Fig. 55 Quantification of percentage of delayed enhancement.

Fig. 56 Cine short axis & vertical long axis views showing asymmetric septal & inferior wall hypertrophy.

Fig. 57 Inflow-outflow cine image showing candle-flame systolic jet across the LVOT, denoting severe stenosis.

Fig. 58 Quantification of percentage of delayed enhancement.

Tables

Table 1 Normal M mode echocardiographic values for neonates
(Kampmann C, Wiethoff CM, Wenzel A, Stolz G, Betancor M, Wippermann CF, Huth RG, Habermehl P, Knuf M, Emschermann T, Stopfkuchen H. Normal values of M mode echocardiographic measurements of more than 2000 healthy infants and children in central Europe. *Heart*. 2000 Jun;83(6):667-72)

Table 2 Normal M mode echocardiographic values from infancy to 18 years
(Kampmann C, Wiethoff CM, Wenzel A, Stolz G, Betancor M, Wippermann CF, Huth RG, Habermehl P, Knuf M, Emschermann T, Stopfkuchen H. Normal values of M mode echocardiographic measurements of more than 2000 healthy infants and children in central Europe. *Heart*. 2000 Jun;83(6):667-72)

Table 3 New York Heart Association (NYHA) Functional Classification (*The Criteria Committee of the New York Heart Association. 1994*)

Table 4 Demographic data

Table 5 Gender distribution among study group

Table 6 Family history of HCM among study group

Table 7 Family history of HCM among study group correlated to PG LVOT & DE

Table 8 NYHA class distribution among study group

Table 9 NYHA class among study group correlated to DE & PG LVOT

Table 10 MRI data

Table 11 Delayed enhancement distribution among study group

Table 12 Comparisons between fibrosis & no-fibrosis groups regarding New York Heart Association Functional Classification (NYHA) & severity of pressure gradient across the left ventricular outflow tract (PG LVOT).

Table 13 Comparisons between fibrosis & no-fibrosis groups were done regarding different demographic & cardiac functions & measurements.

Table 14 Pearson correlation between DE & different demographic & cardiac functions & measurements

Table 15 Echo data

Table 16 Distribution of pressure gradient across the left ventricular out-flow tract (PG LVOT) among study group; SD=standard deviation

Table 17 Unpaired *t* tests comparison between obstructive and non-obstructive groups regarding the severity of NYHA class.

Table 18 Pearson correlation between PG LVOT & different demographic & cardiac functions & measurements

Table 19 Fibrosis detected among study group by echo

Table 20 Pearson correlation between delayed enhancement percentage (DE%) & echo data

Table 21 The sensitivity, specificity & diagnostic accuracy of fibrosis detection by echo (with the delayed enhancement obtained using CMR considered as the reference)

List of abbreviations

AV	Atrio-Ventricular
BSA	Body Surface Area
CI	Cardiac Index
CMR	Cardiovascular Magnetic Resonance
CO	Cardiac Output
DCM	Dilated Cardiomyopathy
DE	Delayed Enhancement
ECG	Electrocardiography
EDV	End Diastolic Volume
EF	Ejection Fraction
ESV	End Systolic Volume
FS	Fractional Shortening
Gd	Gadolinium
Gd-DTPA	Gadolinium-Diethylenetriamine Pentaacetic Acid
HCM	Hypertrophic Cardiomyopathy
HF	Heart Failure
HOCM	Hypertrophic Obstructive Cardiomyopathy
HR	Heart Rate
ICD	Implantable Cardioverter Defibrillator
IR	Inversion-Recovery
IVS	Inter-Ventricular Septum
LAD	Left Anterior Descending
LGE	Late Gadolinium Enhancement
LV EDd	Left Ventricular End-Diastolic diameter
LV ESd	Left Ventricular End-Systolic diameter
LV	Left Ventricle
LVEDVI	Indexed Left Ventricle End Diastolic Volume
LVESVI	Indexed Left Ventricle End Systolic Volume
LVNC	Left Ventricular Non-Compaction
LVSVI	Indexed Left Ventricle Stroke Volume
Max LVWT	Maximum Left Ventricular Wall Thickness
MRI	Magnetic Resonance Imaging
NYHA	New York Heart Association
PC-CMR	Phase Contrast Cardiovascular Magnetic Resonance
PG LVOT	Pressure Gradient across the Left Ventricular Outflow Tract
PPU	Peripheral Pulse Unit
PSIR	Phase Sensitive Image Reconstruction
ROI	Region Of Interest

RV	Right Ventricle
SAM	Systolic Anterior Motion
SCD	Sudden Cardiac Death
SD	Standard Deviation
SPAMM	Spatial Modulation of Magnetization
SSFP	Steady-State Free Precession
SV	Stroke Volume
TI	Inversion Time
VENC	Velocity Encoding

Introduction

Hypertrophic cardiomyopathy (HCM) is the most common inheritable cardiac disorder, with an estimated prevalence of 1:500. It has many complications that may be severe and include arrhythmia and sudden death being the most frequent cause of sudden cardiac death in the young (**Maron 2005**).

Hypertrophic cardiomyopathy is a pathologic condition with an autosomal dominant genetic inheritance in which there is abnormal myocardial thickening without left ventricular dilatation or another identifiable cause such as aortic stenosis or systemic hypertension. Distribution of left ventricular myocardial hypertrophy is variable, with uniform hypertrophy in some cases and focal hypertrophic changes such as septal or apical forms in others (**Cummings et al. 2009**).

A wide range of clinical manifestations are seen, including atypical chest pain, exertional dyspnea related to diastolic dysfunction, and, unfortunately, sudden cardiac arrest or death. At cardiac MR imaging, either diffuse or focal hypertrophic changes can be appreciated, as well as restricted diastolic filling of the left ventricle (**Cummings et al. 2009**).

Techniques which accurately measures left ventricular wall thickness, integrity and perfusion will therefore be necessary to evaluate the effectiveness of treatment strategies (**Maron 2005**).

There are some technical limitation with echocardiography, for example, reliable quantitative delineation of LV wall thickness is dependent on adequate acoustic windows. Also, because the echocardiographic transducer is situated at a fixed point on the anterior

chest wall, cross-sectional images are often unavoidably obtained with obliquity **(Seidman & Seidman 2001)**.

Cardiovascular magnetic resonance imaging has recently emerged as a new noninvasive imaging modality capable of providing high resolution images in any desired plane **(Mahrholdt 2005)**

MRI has proven to be an important tool for the evaluation of patients suspected of having HCM because it can readily diagnose those with phenotypic expression of the disorder and can potentially identify the subset of patients at risk of sudden cardiac death **(Sipola et al. 2005)**.

MRI has the ability to evaluate wall thickness and the distribution of disease better than echocardiography, especially in the anterolateral wall of the left ventricular myocardium **(Rickers et al. 2005)**.

MRI also has the ability to more accurately evaluate left ventricular mass, volumes, and function than echocardiography and to assess for areas of regional wall motion abnormalities, aneurysms, and foci of delayed enhancement **(Hudsmith & Neubauer 2008)**.

The ability to detect areas of myocardial enhancement in hearts with HCM has been used to identify patients who harbor an “arrhythmogenic substrate and that the presence of delayed enhancement and the number of involved segments correlated with the presence of ventricular tachycardia. Which has shown to be increased in patients at higher risk for sudden cardiac death **(Hansen et al. 2007)**.