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Advantages of using Magnetic Resonance in Cardiology

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Cardiovascular Magnetic Resonance (CMR) is a not invasive diagnostic imaging tool for the detection of the most common heart diseases. It creates images from atomic nuclei with uneven spin using radiowaves in the presence of a magnetic field. For clinical purposes, magnetic resonance (MR) is performed using hydrogen-1, which is abundant in water and fat. Radiofrequency waves excite the area of interest to create tissue magnetization that decays (relaxation) and after a short period is induced to release energy as a radio signal. These echoes are converted with Fourier transformation into images of spatially resolved radio signals. Relaxation is quantified in spatially orthogonal directions as T1 and T2, which allows tissue characterization to serve as a powerful clinical tool. Recently CMR has become the gold standard for evaluating myocardial function, volumes, and scarring. It is an indispensable tool in the evaluation of congenital heart disease, heart failure, cardiac masses, pericardial disease, and coronary artery disease. Cardiovascular magnetic resonance imaging is unique in its comprehensive tissue characterization, including assessment of myocardial oedema, myocardial siderosis, myocardial perfusion, and diffuse myocardial fibrosis. This review is focused on advantages of the detection of suspected cardiomyopathies, in particular in the case of the dilatative cardiomyopathy, the hypertrophic cardiomyopathy and the right ventricular arrhythmia cardiomyopathy that have been examined in this tractation

Keywords: Cardiovascular Magnetic Resonance, Cardiac Imaging, Heart Diseases, Cardiomyopathies, Hearth Failure

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Introduction

In a few years magnetic resonance imaging has taken a crucial role in the study of cardiomyopathy both on an ischemic and non-ischemic basis, and in the evaluation of patients with heart failure¹⁵. This technique, known as Cardiovascular Magnetic Resonance, is unique to the diagnosis and assessment of cardiomyopathies. Consequently, in cardiomyopathy magnetic resonance is today a precise and reliable instrument for diagnosis and for prognostic stratification when it comes to a structural damage⁶.

What can be diagnosed by using the cardiac magnetic resonance imaging?

Due to the development of specific image sequences, the use of a medical contrast medium and the implementation of innovative solutions such as hardware and image reconstruction, it is now possible to check heart anatomy and function. In particular, physicians can investigate heart perfusion, oedema, fibrosis, haemorrhage, and iron content of the cardiac muscle².

The classic Cardiovascular Magnetic Resonance imaging (CMR) sometimes known as cardiac MRI scanner prototype - provides a magnetic range of 1.5-3T, a dedicated coil for multi-channel heart study, and specific software that includes sequences for heart study, as well as appropriate calculation algorithms⁵. Today all scanners can be used for heart and vase studies as long as hardware and software for electrocardiogram signalling are available. These are necessary to synchronize the data acquisition with the movements of the heart, and the sequences suitable for the evaluation of both morpho-functional and variations of the signals induced by the blood flow and contrast media¹⁸. The standard exam begins with dark blood sequences for classical anatomical evaluation, followed by several light blood sequences for the volume and kinetic analysis of cardiac chambers and valve devices. The use of a contrast medium – the most widely used are gadolinium chelates - influences the magnetic properties of the myocardium. Analysis of the signal intensity variations over time at the first passage allows evaluation of any perfusion heterogeneity, while late-on analysis allows verification of the presence of extra vascular accumulation, usually associated with fibrosis²⁻²⁶.

The CMR can also highlight areas of delayed enhancement in cardiomyopathy, and it is now clear that this phenomenon is linked to the particular tissue and cellular distribution of gadolinium chelate7. The delayed enhancement phenomenon occurs when, a few minutes after the administration of the ferromagnetic contrast medium, the signal increases over the surrounding myocardium because of the

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changes in magnetization induced in the tissue under examination. In the case of acute infarction, gadolinium penetrates the cells and produces the typical signal increase at the delayed enhancement after a few minutes of the lesion. In the later phase, known as the repair phase, gadolinium occupies the extracellular space left free from the cells. Thus, the presence of delayed enhancement areas simply reflects the repairing fibrosis. A complete study of the heart anatomy, function, rest and stress perfusion, as well as delayed enhancement, can be completed in 40-50 min. The absence of ionizing radiation and the good quantity of information that can be obtained in one exam, make the CMR a technique of great clinical interest²⁻²⁶.

Cardiomyopathies

Cardiomyopathies (CMPs) are very different myocardial diseases. According to the classification of the American Heart Association in 2006, they are distinguished by primitive forms and secondary forms. Among the primitive forms are those acquired, genetically transmitted, and a mixture of the two. The European Cardiology Society in 2008 differentiated cardiomyopathies on both morphological and functional aspects.

The CMR can characterize the clear majority of CMPs of clinical interest from the morpho-functional point of view. No single diagnostic tool has the versatility, robustness, simplicity, immediacy of information, accuracy, and clinical applicability of CMR.

In a recent CMR consent document, the use of CMPs is considered appropriate to both identify an aetiology of a cardiopathy due to an unknown cause associated with heart failure, and to better assess patients with suspected but not characterized cardiopathies10. For example, the ischemic aetiology or related coronary atherosclerosis are generally easy to detect due to the presence of one or more

lesions of the left ventricle that characterize it11-13. Today CMR is the only non-invasive method that can detect a precocious infarction with high accuracy. Outside the ischemic aetiology, CMR can identify the aetiology of a cardiopathy in the presence of a left ventricular dysfunction of uncertain origin¹²⁻¹⁵.

Thanks to the combination of morpho-functional aspects and to the presence of specific delayed enhancement pattern (observed after the administration of the contrast medium), the CMR can detect with a high degree of accuracy: primitive dilated cardiomyopathy, hypertrophic cardiomyopathy, amyloidosis and other forms of restriction, right ventricular cardiomyopathy, and other diseases that may result in non-ischemic myocardial injury.

Over the years CMR has not only played a major role in the diagnosis of cardiopathy but has also shown significant prognostic ability6. The prognostic role of CMR is demonstrated by its ability to identify the presence and to measure the extent of myocardial scarring. CMR can work with recognized markers and factors usually detected with the other relevant diagnostic methods¹⁴.

The presence and magnitude of myocardial fibrosis is particularly relevant, not only because it is traditionally evaluated, but because it makes up a large part of the clinical research. New sequences and specific technical approaches for the collection of quantitative and accurate measurements of pathological quantities of collagen, allows CMR to be used for both diagnosis and prognosis of in many pathological conditions including cardiomyopathies¹⁶.

In conclusion, for all the above-mentioned features, it can be stated that CMR can be considered a definitive examination technique to identify both aetiology and prognosis in the presence of many established or suspected cardiomyopathies.

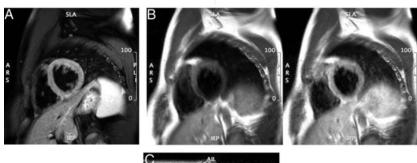




Figure 1: Cardiovascular Magnetic Resonance of Acute Myocarditis

Panel A: T2-weighted image of LV myocardial edema showing global bright signal intensity (ratio 2.2) of the left ventricle relative to the myocardium.

Panel B: Early enhancement (T1-weighted spin echo) before (left) and after (right) Gd administration; enhancement ratio 5.4.

Panel C: Arrows indicating late enhancement (T1-weighted gradient echo sequence with myocardial nulling) 10 minutes after Gd. Gd indicates gadolinium; and LV, left ventricular. Hundley W.G., Bluemke D.A., Finn J.P. et al. ACCF/ACR/AHA/NASCI/SCMR 2010 Expert Consensus Document on Cardiovascular Magnetic Resonance 2010, 55, 2614-2662

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The dilatative cardiomyopathy

In the primitive dilative form, CMR provides for the evaluation of volumetric, mass and ventricular kinetic parameters that are not affected by geometric changes in the heart. These parameters can be measured accurately, and this allows surgeons to evaluate whether an implanted device is required. In addition, it provides accurate prognostic elements and highlights opportunities for unplanned prognostic stratification¹².

The most interesting element of CMR imaging is the ability to exclude the ischemic aetiology of a cardiomyopathy with a high degree of accuracy. This is usually the result of one or more myocardial necrosis that causes scars that can be identified using the delayed enhancement technique. In the so-called "primitive" forms, it is possible to document predominantly intra-myocardial scars. However, sometimes scars appear

"ischemic" 12-15 and so the absence of subendocardial or transmural delayed enhancement areas can be used to virtually exclude the presence of ischemic heart disease 17,18.

CMR diagnostic targets in dilatative forms are: progressive left ventricular dilatation, changes in systolic function, and the presence of myocardial fibrosis. Myocardial fibrosis is correlated to the presence of malignant ventricular arrhythmias and, consequently, there is a probability of effective discharge of implantable defibrillators used in primary prevention¹⁹.

For several years, CMR has been shown to play a major role in recognizing signs of inflammation and myocardial involvement in myocarditis²⁰. It has also shown that it can highlight, locate, and quantify both oedema and hyperaemia as well as the fibrosis that occurs in this condition. Today, it is also possible to diagnose myocarditis in 80% of cases by using specific sequences with CMR alone²¹.

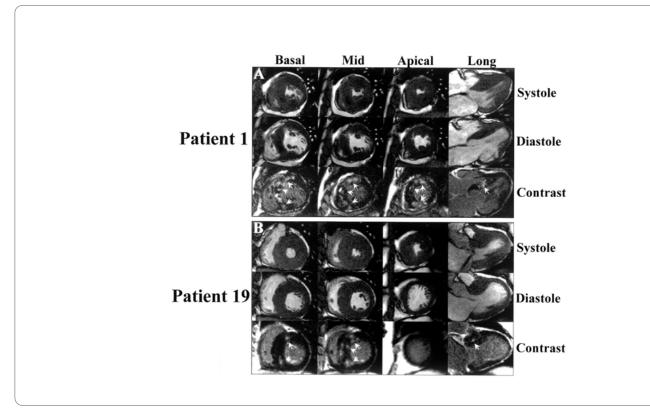
Pathology expression is determined by oedema, hyperaemia and cellularity that participate in inflammation, which is manifested by the typical but not exclusive side of the lateral wall, the predominant subepicardic expression, and often the complete transitionality of delayed enhancement. Such features may, in selected cases, allow diagnosis, while avoiding invasive manoeuvres such as endomyocardial biopsy^{21,22}.

The hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy is generally easily recognized by echocardiography and does not pose significant problems with pathophysiological assessment. By using the standard echocardiographic examination, the presence and magnitude of an obstruction, involvement of the left atrium, or any non-synergic areas can be easily highlighted. CMR over the years has demonstrated superior ability to recognize hypertrophy areas also present in myocardial segments that are otherwise unobtrusive^{23,24}. With the use of delayed enhancement for several years it has been possible to document the presence of scar tract areas in the left ventricle of many patients with hypertrophic cardiomyopathy²⁵.

The recent American College of Cardiology Foundation (ACCF)/AHA guidelines for the diagnosis and treatment of hypertrophic cardiomyopathy have introduced CMR for the first time among techniques that have a major impact both in the differential diagnosis of this pathology and in prognostic stratification. This gives CMR a key role in detecting areas of ventricular scarring²⁶.

Finally, CMR was used in Fabry's disease of chromosome X linked lysosomal disease caused by the deficiency of the alpha-galactosidase enzyme. In a recent register, 0.5% of patients with suspected hypertrophic cardiomyopathy have this nosological entity²⁷



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Figure 2: Representative patient images.

(A) Patient with asymmetric septal hypertrophy, maximum wall thickness of

20 mm, normal ejection fraction, and marked myocardial scarring (hyperenhancement shown by arrows).

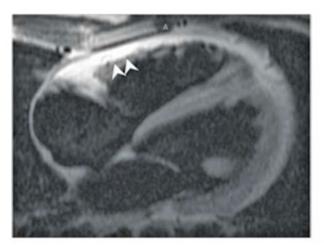
(B) Patient with greater hypertrophy (maximum wall thickness, 27 mm) but with less scarring. In both patients, there are multiple foci of scar, which are predominantly mid-myocardial in location and are not present in the lateral free wall. The long- axis cines images of Patient 19 demonstrate systolic anterior motion of the mitral valve, systolic flow turbulence in the left ventricular outflow tract, and mitral regurgitation. Choudhury L., Mahrholdt H., Wagner A., et al. Myocardial

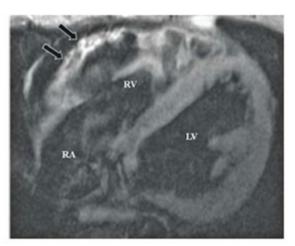
scarring in asymptomatic or mildly symptomatic patients with hypertrophic cardiomyopathy 2002, 40, 2156-64

The right ventricular arrhythmia cardiomyopathy

The first description in the literature of a diagnosis made by CMR was in 1987 and featured a case of right ventricular arrhythmia cardiomyopathy (CAVD)²⁸. Since then, much progress has been made and today CMR can highlight morpho-functional alterations more accurately than those obtained by echocardiography. Specifically, improvements have been made to the alterations of the endocardial profile, diastolic bulging areas, microaneurisms, and accentuated trabecularity. Similarly, changes in the volume and kinetic of the right ventricle are easily detected²⁹³¹. Today CMR is considered the most

reliable and accurate technique for the study of the right ventricle³². CMR can highlight areas of reduced systolic wall thickening and areas of altered regional kinetics, which may also affect the left ventricle in advanced cases. Along with the alterations of the regional and global kinetics of the right ventricle, and in the absence of known causes, patients with CAVD may exhibit alterations in the wall signal after administration of the contrast medium³³. The ACC / AHA attributed the highest degree of appropriateness for this application, and this was also confirmed by a review of the diagnostic criteria of the disease by a special Commission of the European Cardiology Society³⁴.





Calkins H,et al. In Shenasa M, et al, editors. Cardiac Mapping. 4th edition. New York: Wiley-Blackwell; 2013. p. 680;2013.

Figure 3: MRI in a patient with arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVD/C) Calkins H. et al. In Shenasa M. et al, The future of cardiac mapping: Dawn of a New Decade, in Cardiac Mapping, M Shenasa, Editor. 2013

The other cardiomyopathies

Over the last few years, all major cardiomyopathies have been examined with CMR, which has allowed their main features to be described. Among these there are: non-compaction cardiomyopathy, heart amyloidosis, and numerous other primitive or secondary myocardial conditions such as sarcoidosis or hemochromatosis^{35,36}.

Differential diagnosis of cardiomyopathies

The clinical diagnosis of non-ischemic cardiomyopathy is often linked to the exclusion of a diagnosis of coronary artery disease. Consequently, the standard exam used to exclude coronary heart disease is coronary angiography. This examination offers a luminous view of the coronary artery vessels, allowing the presence of coronary obstructive lesions to be accurately highlight. Today it is clear that

conventional coronary artery disease is not able to accurately describe the presence or absence of non-obstructive coronary atherosclerosis. This can be more effectively described by intravascular ultrasound or coronary computerized tomography^{38,39}. Thanks to the increasing use of coronary multilayer computerized tomography, it is now possible to exclude coronary heart disease in the presence of an unknown ventricular dysfunction in all patients⁴⁰.

Conclusions

CMR is a non-invasive, polyparametric, quantitative, repeatable diagnostic method that is particularly useful in the diagnosis, follow up, and prognostic stratification of patients with cardiomyopathy. Within the limits of etiologic classifications, CMR can recognize most cardiomyopathies through identification of peculiar features based on anatomical, functional, and structural aspects that would otherwise

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be unavailable. In some circumstances it can be a valid alternative to myocardial biopsy, especially where the CMR diagnostic aspects are particularly specific. In many cases CMR can also detect the aetiology of cardiomyopathy and optimize the clinical pathways through an otherwise unpredictable prognostic evaluation. Despite the crucial role of other diagnostic methods, primarily echocardiography, CMR is today a tool that allows unprecedented level of clinical vision both on the diagnostic side and in the therapeutic choice. In addition, it can also identify the risk of patient emergencies with suspected or confirmed cardiomyopathy with an increased degree of accuracy.

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