

Cardiovascular Magnetic Resonance Imaging in the Evaluation of the Right Ventricle and its Pathologies

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Abstract

Cardiac magnetic resonance imaging (CMRI) is a non-invasive three-dimensional method that allows morphological and functional evaluation of all chambers of the heart. This method has gained importance in the identification and characterization of primary diseases involving the right ventricle (RV), and secondary diseases that lead to dysfunction of the right chambers. This study aims thereby to elucidate the best methods to diagnose these diseases by means of the Cardiovascular MRI.

Introduction

Normally, the right ventricle is anteriorly positioned to the left ventricle (LV), right behind the sternum, and delimited by the annuli of the tricuspid valve and the outflow tract, which communicates with the pulmonary¹ Unlike the left ventricle (LV), which has a symmetric ellipsoidal morphology, RV shape is complex, triangular when viewed in profile, and has a crescent shape when viewed in cross-section.^{2,3} Such structure has the function to house the whole venous system returning to the heart and is therefore vulnerable to any sharp increase in wall stress.⁴

Cardiac magnetic resonance imaging (CMRI) has gained importance in viewing and evaluating the RV, as it allows to explore its anatomy and function in a noninvasive, three-dimensional manner, and free of limitations in relation to the patient's chest, in addition to dispensing with radiation exposure. It also performs an outstanding cardiac tissue characterization and accurately quantifies ventricular volumes and valve regurgitation, in addition to allowing the analysis of adjacent vascular structures (pulmonary veins and arteries), thus making this imaging method preferable over other examinations.⁵⁻⁷ Understanding the RV remodeling process is

important to better understand the response of this chamber to the increases in pressure and volume, and thereby correlate these changes with symptoms shown by the patient, resulting in better diagnostic and therapeutic delineation.⁸ Thus, during RV analysis, it should be kept in mind the diseases that can lead to secondary changes in that cavity, as well as primary cardiac diseases involving the right chambers (Table 1).

This study aims to describe the most appropriate methodology for a better diagnostic clarification in patients with right ventricular dysfunction, and expose the most common primary and secondary cardiac diseases that can affect the RV, and CMR findings in each of them.

CMR Imaging Protocols

For RV to be morphologically and functionally evaluated, sequences of steady state free precession (SSFP) cinemagnetic resonance (Cine-MRI) should be primarily performed, in previously determined sections (4-chamber, short axis view of the entire cavity, and longitudinal plane of RV or RV outflow tract, which include RV outflow infundibulum and outflow tract). These are the basic recommendations for any examination, according to protocol guidelines of the American Society for Cardiovascular Magnetic Resonance (SCMR).⁹

From these sequences and at the moment of acquisition, anomalies in the RV structures should be identified by an experienced examiner, on the basis of visual analysis, such as increased cavity, tapered, akinetic regions of the wall, with microaneurysms, trabecular hypertrophy beyond evaluation function, whose quantification should be performed later in a semi-quantitative manner. It is also important to look for changes in transvalvular jets indicating stenosis or regurgitation. In the presence of these, they should be quantified by means of flow analysis (phase contrast) of the pulmonary artery trunk. Where intracavitary shunts are identified, this sequence should be carried out also in the aortic root, so that the presence of hemodynamic repercussion passing through the flow ratio between the aorta and pulmonary artery (Qp/Qs) can be identified.

In order to complement these sequences in case of anomalies in right cavities in which there is not yet known any diagnosis at the moment of acquisition, it is recommended that evaluation of the pulmonary vessels be performed by means of the Cine-MRI. This sequence is used in the axial plane of the chest, totally covering the heart area, including trunk of pulmonary artery and its branches (10 to 12 sections of 10 mm and 0 mm gap, with 300/400-mm FOV and matrix 150-200 mm, with only five phases of the cardiac cycle), and it can be performed with one or two breathing pauses only.

Keywords

Magnetic Resonance Angiography; Right Ventricular Dysfunction; Right Ventricular Arrhythmogenic Dysplasia; Diagnosis.

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Table 1 – Primary and secondary heart diseases that involve the right ventricle and result in morphological and functional changes in that chamber

Primary RV diseases (congenital and acquired)	Secondary RV diseases
- Arrhythmogenic right ventricular dysplasia	- Pulmonary hypertension a) primary b) secondary b.1) LV dysfunction b.2) pulmonary embolism b.3) valve insufficiency of the left ventricle
- RV endomyocardial fibrosis	- Volume overload a) Anomalous drainage of pulmonary veins b) Left-to-right shunts c) Valve insufficiencies
- Ebstein Anomaly	- Pressure overload a) Pulmonary Stenosis b) Outflow tract obstruction caused by masses
- Tetralogy of Fallot	- RV Infarction

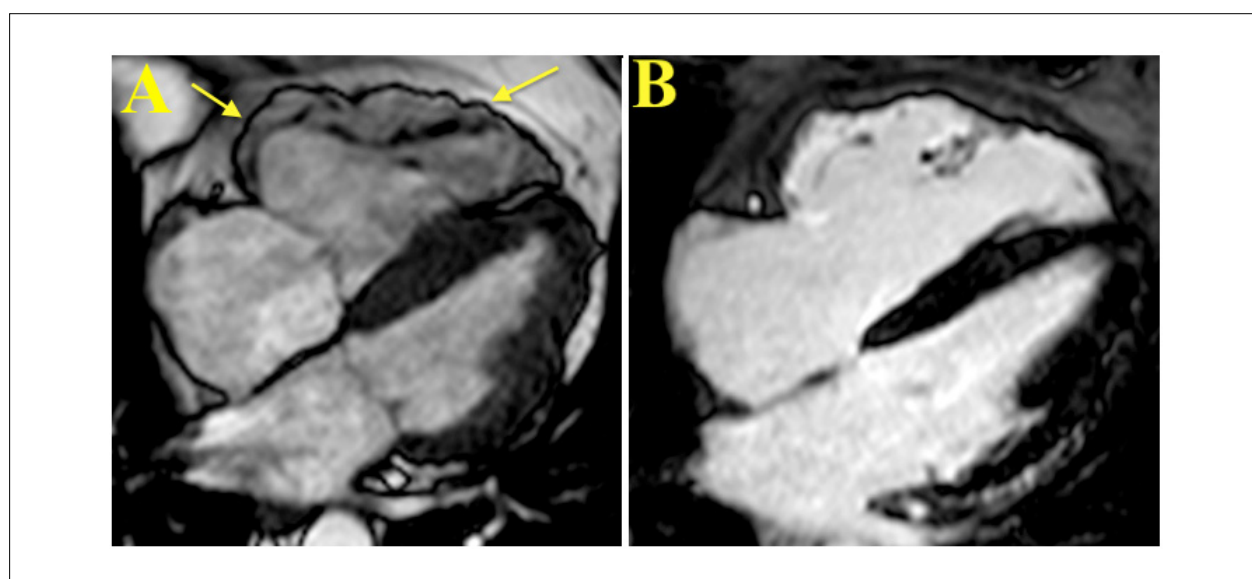


Figure 1 – 55 year-old male patient diagnosed with ARVD, after observation of increased right chambers and RV systolic dysfunction. A: RV “Accordion sign” and formation of microaneurysms (arrows) can be observed; B: Late enhancement MRI analysis after gadolinium injection showing predominant fibrotic areas in the RV free wall.

This will allow for the analysis of the pattern of pulmonary drainage and major branches of the pulmonary arteries. In case of any anomalies of these structures, a Gadolinium-based contrast magnetic resonance angiography (MRA) of the vascular territory in question (pulmonary artery or veins) is performed to verify the suspected anomaly.

Finally, 10 minutes after contrast injection, the late enhancement MRI sequence is performed, which helps diagnose specific diseases, such as RV infarction and arrhythmogenic right ventricular dysplasia (ARVD).^{10,11} The importance of this technique lies in delimiting areas of necrosis and fibrotic tissue in the myocardium by increasing the intensity of tissues and decreasing intensity of the signal from the myocardium.¹²

Some cases extracted from our work will help us illustrate the role of CMRI in the evaluation of RV and the importance of a comprehensive approach to cardiac and vascular structures. All of the images shown here were obtained from tests performed at the Hospital e Fundação São Lucas’ MRI Service, located in the city of Aracaju, State of Sergipe, Brazil, in which all patients underwent a high field cardiovascular magnetic resonance (using 1.5T Magneto Phillips AchievaTM) from 2011 to 2015.

Arrhythmogenic right ventricular dysplasia diagnosis

The arrhythmogenic right ventricular dysplasia (ARVD) is an inherited and rare cardiomyopathy characterized by a progressive replacement of right ventricular myocardium

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with scar tissue, leading to progressive dysfunction of this heart chamber.¹³ This disease is estimated to be responsible for 10% of sudden deaths in individuals less than 35 years of age.¹⁴ Its progression starts at the epicardium and the middle layer of the myocardium, then propagating transmurally. The evolution of the condition results in the wall thinning and formation of aneurysms typically located in the RV lower, apical and infundibular walls.¹⁵ Clinical manifestations chiefly include ventricular tachycardia, with bundle of His left branch block morphology and sudden death.¹⁴⁻¹⁶

The accuracy of the CMRI examination in the characterization of these changes in the RV and the study of its function makes this the method of choice for evaluation of ARVD, rendering an excellent diagnostic accuracy in addition to better reproducibility among several experienced observers.^{16,17} Tandri et al.¹⁷ demonstrated sensitivity of 75% and specificity of 97% in the use of CRMI to study abnormalities in the regional movement of RV walls, with a resulting positive predictive value of 90% in the study of the disease.

The ARVD diagnosis by means of CMRI should be done by identifying RV dilation and systolic dysfunction of this chamber

by using Cine-MRI sequences, and the presence of dyskinetic areas with formation of microaneurysms, according to published and revised criteria (*task force criteria*).¹⁸ An important finding in the CMRI study was described by Dalal et al.,¹⁹ who detected “wrinkling” of the free wall, in the right ventricular outflow tract and subtricuspid area, more prominent in systole and not affecting the overall function of this chamber, referred to as “Accordion Sign” by the authors. Finally, after a late enhancement analysis, fibrosis areas are commonly found in this wall, which has been associated with a higher incidence of ventricular tachyarrhythmia and hence higher mortality rates.^{17,20}

Tricuspid insufficiency diagnosis

Changes in the right valves, especially in the tricuspid valves, are commonly undervalued in everyday clinical practice due to the greater importance of the left side of the heart in valvular heart diseases.²¹ The study by Framingham,²² however, detected signs of tricuspid insufficiency (TI) in 82% of the male population and 85.7% of the female population, examined by means of transthoracic Doppler echocardiography (TTD)

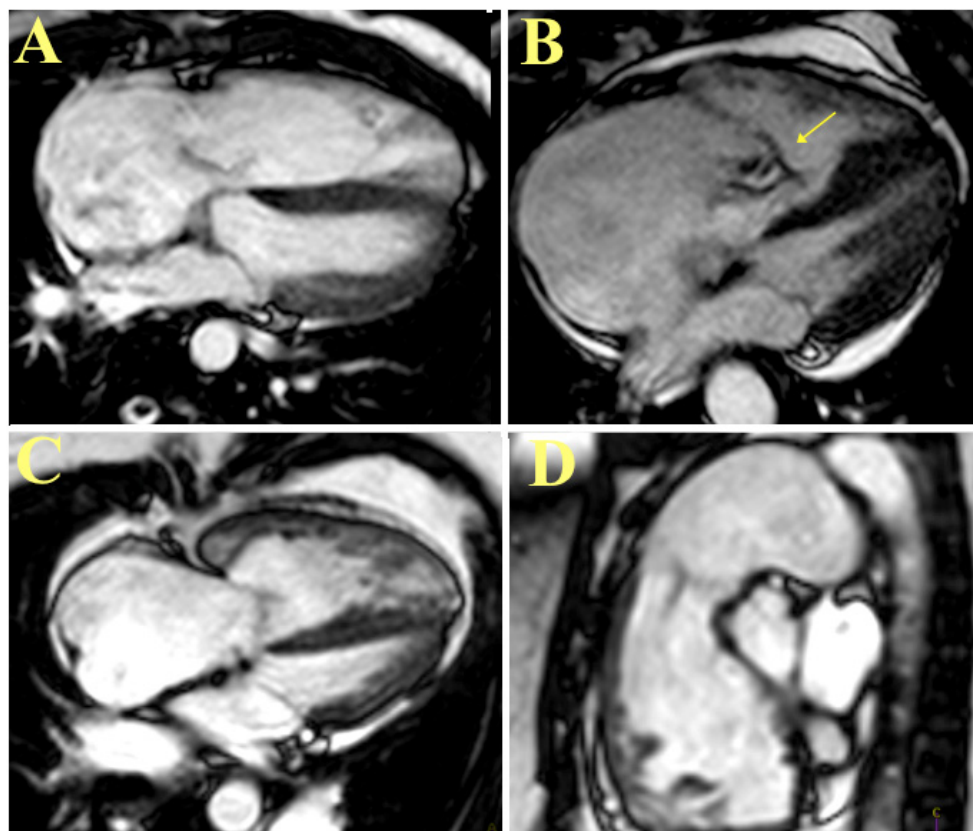


Figure 2 – Morphological and functional analysis of the heart by means of SSFP Cine-MRI in patients with dilated right heart chambers. A: presence of a swirling flow along the cavity resulting from severe tricuspid regurgitation (regurgitant fraction calculated using phase contrast = 76%). B: Ebstein's anomaly with tricuspid valve implantation 3 cm below the mitral valve plane (arrow). C and D: Visible Primary PH diagnosis, and increased right chambers on Cine-MRI (C), right branch dilation upon evaluation of pulmonary arteries (D).

method. Moreover, Nath et al.²³ have shown that a greater reflux volume in TI is commonly associated with RV dilation and dysfunction, and higher atrial pressure.

The TTD is still the main imaging method in the initial evaluation of patients with valvular changes. However, the CMRI importance in studies is growing thanks to the possibility of evaluating the entire heart and all of its 4 valves without restrictions such as body mass and coexisting lung diseases.²⁴

The TI study consists of a preliminary morphological and functional analysis of the right chambers by using steady-state free-precession (SSFP) cine MRI, allowing the evaluation of any increase in these structures as a result of volume overload generated by valvular regurgitation. If other secondary diseases that would justify the morphological and functional changes in RV are not detected, this suggests that it is related to the primary dysfunction of the tricuspid valve. For evaluation and quantification of reflux, the stroke volume is calculated by using Simpson's technique,²⁵ then, the result is subtracted from the actual contrast volume through the LV outflow tract, obtained by using the phase contrast technique.¹²

Ebstein's Anomaly diagnosis

The Ebstein's anomaly is characterized by a failure of delamination of the tricuspid valve in the embryonic stage, culminating with variable degrees of apical displacement of the septal and posterior leaflets.²⁶ How this apical displacement and the resulting valve regurgitation affect RV size and operation is still the subject of discussion. The CMRI 4-chamber view enables the analysis of the insertion of the tricuspid leaflets and their points of coaptation, allowing for more detailed delineation of the functional components of the RV and better reproducibility in the study of this malformation.²⁷

Moreover, real-time visualization of the heart through Cine-MRI technique allows for delineation of morphological

and functional changes of the right chambers resulting from anatomical malformation. It is also possible to carry out a phase contrast sequence for analysis of the valve flow and identification of concomitant valvular regurgitation.

Primary pulmonary hypertension

Dresdale et al.,²⁸ in the 1950s, were the first to describe hypertensive vasculopathy in the pulmonary circulation detected by catheterization. According to the authors, this disease, which they referred to as primary pulmonary hypertension (PH), was characterized by vasoconstriction and increased pulmonary artery pressures, with no apparent cause of mitral stenosis or pulmonary parenchymal disease. Primary PH is a rare condition, with a high mortality rate, even after the development of various treatment strategies.²⁹

The classic method for diagnosis of PH is right heart catheterization with a mean pulmonary artery pressure > 25 mmHg and pulmonary artery occlusion pressure <15 mmHg.³⁰ The progressive increase in pulmonary artery pressure and the pulmonary vascular resistance result in right ventricular overload, with hypertrophy, dilation and consequent dysfunction of this chamber.^{30,31} CMRI provides important information about changes in the right ventricular function and morphology, such as increased thickness of its wall and paradoxical movement of the interventricular septum, as well as dilation of pulmonary arteries, important features in the diagnosis of primary PH, as well as to rule out other diseases that can lead to changes in this cavity.³⁰⁻³² Late enhancement frequently occurs at the insertion points of the right ventricular free wall to the interventricular septum, in patients in advanced stages of the disease, and is related to significant impairment in the function of the ventricular cavity.³³ As already shown in the literature, reduced RV function detected in the CMRI is considered an important predictor of increased morbidity and mortality in patients with PH.³⁴

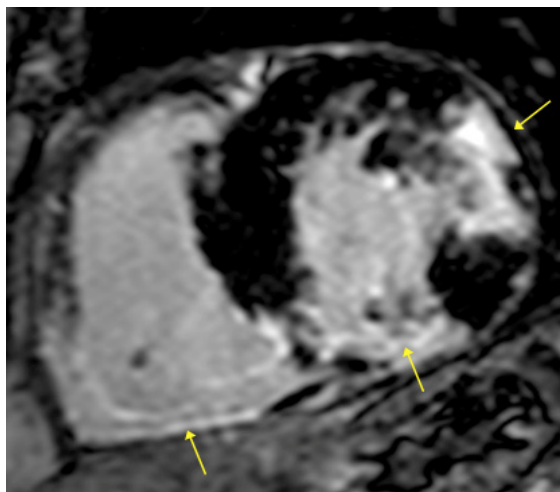


Figure 3 – 73 year-old male patient, in whom late enhancement areas (arrows) are viewed using CMRI, after gadolinium injection, which are related to transmurular infarction of the inferior, inferoseptal, anterolateral walls of the LV, the latter extending towards the RV inferior and free walls.

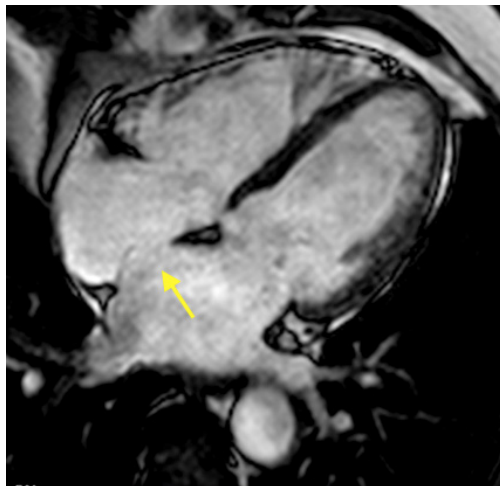


Figure 4 – 61 year-old male patient, with atrial septal defect on Cine-MRI sequences (arrow), and Qp/Qs ratio = 1.5 calculated after an image sequence in phase contrast. A slight increase in right chambers and severe increase in the left atrium are observed.

RV infarction diagnosis

Although isolated RV infarction is extremely uncommon, it may be present in about one-third to half of patients with left ventricular (LV) inferior wall myocardial infarction, generally identified by ST segment elevation at inferior and anterior leads.³⁵ However, when compared to the electrocardiogram (ECG) and TTD, CMRI has greater sensitivity in the detection of these regions at the right ventricular cavity.³⁶

Evaluation of infarcted areas in the RV using CMRI is carried out using late enhancement, an already established method for the diagnosis of infarction.³⁷ Its importance lies in that it allows for the finding of infarcted myocardium in the LV and along its extension towards the RV wall.¹¹ Morphological and functional assessments should also be carried out for quantification of right ventricular function.

Atrial septal defect assessment

Defects in the atrial septum are responsible for approximately one third of cases of congenital heart disease in adults. Patients with moderate or severe atrial septal defects (ASD) may have increased pulmonary blood flow with high pressure in the pulmonary artery, resulting in a shunt reversal due to the Eisenmenger syndrome.³⁷

Transesophageal Doppler echocardiography is the most widely used imaging technique for the diagnosis and quantification of atrial septal defects. CMRI, however, by means of morphological evaluation in SSFP sequence and flow evaluation (phase contrast), has been proven to be complementary to the echocardiographic examination in situations where it is necessary to avoid the more invasive

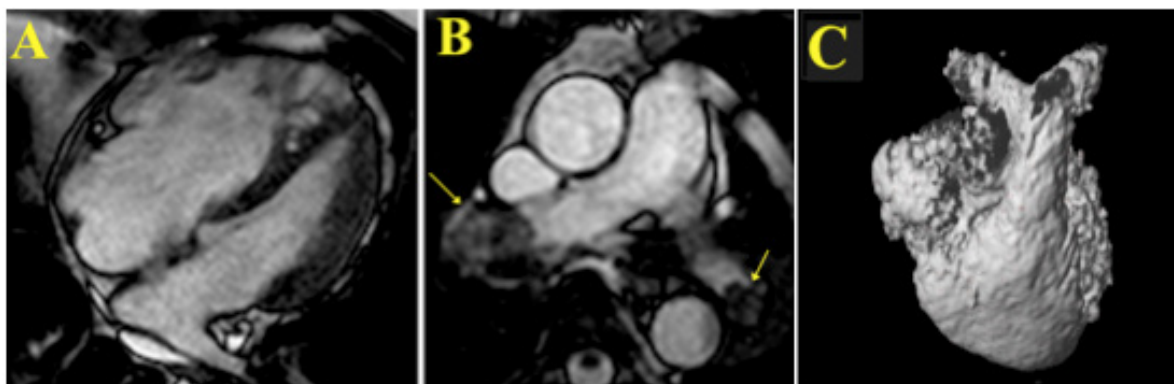


Figure 5 – A 59-year-old male patient, whose cine-MRI shows increased RV associated with dysfunction of the chamber (A). These changes were justified by the finding of thrombi (arrows) in the left and right pulmonary arteries (B), and interruption of the pulmonary arteries bilaterally on MR angiography (C).

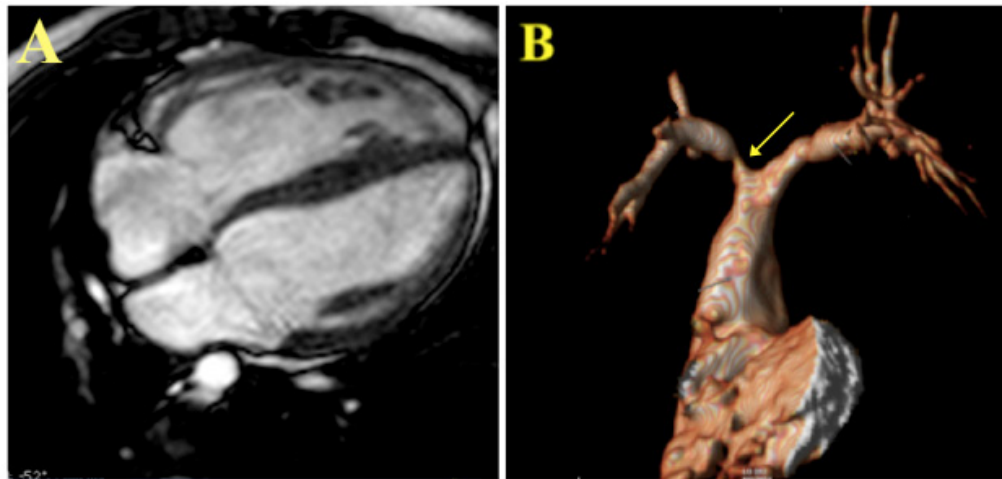


Figure 6 – A 17-year old male patient, after tetralogy of Fallot correction surgery. It can be observed an increase in RV cavity and in the thickness of its walls on cine-MRI (A), as well as severe luminal reduction in the origin of the pulmonary artery using MR angiography (B - arrow).

method.^{38,39} By measuring the pulmonary artery and ascending aorta blood flow, the Qp/Qs ratio is found, and when its result is greater than 1.5, hemodynamic repercussion is assumed to exist, thus indicating surgical correction.⁴⁰

Pulmonary thromboembolism diagnosis

Pulmonary embolism occurs as a consequence of a thrombus, mostly formed in the deep venous system, which dislodges and travels through the right chambers of the heart, then blocks one of the pulmonary arteries or their branches, thus characterizing the venous thromboembolic disease or pulmonary thromboembolism (PTE).⁴¹ Its annual incidence is estimated to be 60-70 cases per 100,000 inhabitants, with a mortality rate four times higher when treatment is not established.^{42,43} This aggravates the great risk to the affected patient's life due to the difficulty in completing the clinical diagnosis as a result of the non-specific aspect of its signs and symptoms.⁴⁴

The RV undergoes significant morphological and functional changes after PTE, resulting from the increase in its afterload, and may be present in the early stages of the disease. The study of its function using CMRI SSFP sequences is an important prognostic marker.⁴⁵ Moreover, in case of suspected acute or chronic PTE, the examination should be complemented by MR angiography of the pulmonary arteries, which allows more accurate identification of filling defects of the right and left pulmonary arteries. This technique has become an alternative to other methods that use ionizing radiation and an iodinated contrast agent.⁴⁶ However, CT angiography is still the most accurate examination for evaluation of subsegmental vessels.

Cardiac evaluation after correction of tetralogy of Fallot

Tetralogy of Fallot (TF), characterized by pulmonary stenosis, dextroposition of the aorta on the interventricular

septum, ventricular septal defect and right ventricular hypertrophy, is the most common cyanotic congenital heart disease. Patients usually have good results after total surgical correction, although they can have hemodynamic changes in the postoperative period, the most common being RV dilation resulting from pulmonary regurgitation, atrial and/or ventricular septal defects, tricuspid regurgitation, aneurysm of the RV outflow tract, pulmonary artery stenosis, tachyarrhythmias.⁴⁷⁻⁴⁹

CMRI is important in the preoperative evaluation of this disease as it allows for better delineation of both cardiac and vascular morphology. However, its major role is in the TF post-repair monitoring. In addition to the morphological analysis, RV and LV functional analysis is performed using SSFP cine-MRI sequences, in long and short axis view. Furthermore, the use of late enhancement makes it possible to observe myocardial fibrosis points, which generally are associated with left ventricular dysfunction, exercise intolerance, and changes in regional contraction.⁵⁰

Flow analysis using phase contrast allows to quantify pulmonary insufficiency, which commonly results from the disruption of the integrity of this valve for relieving the obstruction of the RV outflow tract.⁴⁹ This methodology allows quantification of possible tricuspid regurgitation. In some cases, the MR angiography analysis of pulmonary arteries is valid. The importance of this analysis was highlighted by Sheikh et al.,⁵¹ who showed that 18.92% of the total number of patients who had undergone surgery for TF correction had abnormalities in the pulmonary artery after catheterization.

Diagnosis of Anomalous drainage of pulmonary veins

Partial anomalous pulmonary venous drainage (APVD) is a rare congenital anomaly in which one to three pulmonary veins drain into the right atrium or one of its tributaries,

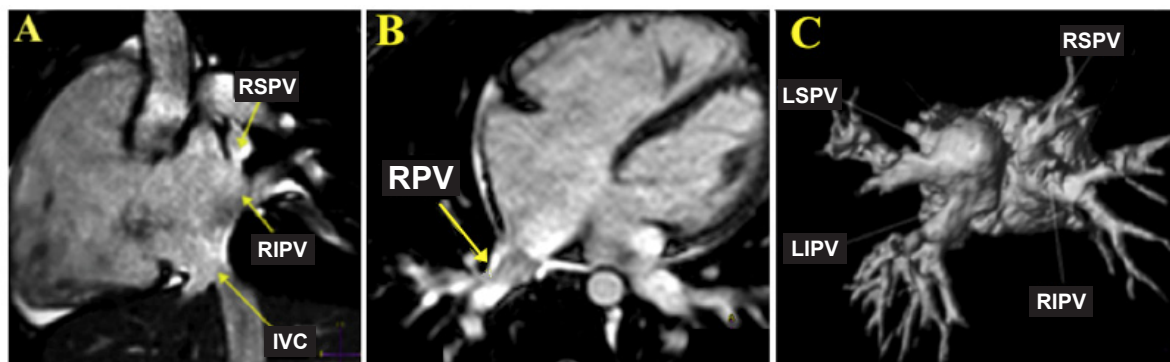


Figure 7 – A 22-year-old female patient being examined by CMRI, whereby an increase in right cavities with preserved ventricular function was observed. A and B: Cine-MRI sequence revealing right pulmonary veins (RPV), right superior pulmonary veins (RSPV) and right inferior pulmonary veins (RIPV) and right inferior pulmonary veins (RIPV) flowing into the right atrium, in addition to the inferior vena cava (IVC). C: MR angiography of the pulmonary veins in a top view, confirming the presence of partial anomalous pulmonary venous drainage, with two pulmonary veins flowing into each of the atria. LSPV: left superior pulmonary vein; LIPV: left inferior pulmonary vein.

instead of going to the left atrium.⁵² TTD has been the noninvasive method more commonly used in the evaluation of the pulmonary venous system. However, the sensitivity of this technique is limited due to acoustic windows, often making it necessary to use the transesophageal method. Should diagnostic uncertainty persist, imaging methods with three-dimensional reconstruction (CMRI and tomography) allow the study of the pulmonary veins in more detail, by following their trajectory from the periphery to its points of entry in the heart.⁵³ CMRI allows, by using the Cine-MRI sequence in axial plane of the chest, to identify abnormalities in pulmonary venous drainage into the atria, and such finding should be confirmed, preferably at the time of the study, by using MRA.⁵²⁻⁵⁴

Conclusion

RV CMRI evaluation has advantages over other methods available, due to both detailed analysis of its morphology and function and the help it provides in the etiologic elucidation of primary or secondary diseases. It also provides data related to hemodynamic repercussions and tissue changes, which are fundamental for prognostic evaluation and management of these diseases.

In order for such help to be provided, a protocol for a complete, supervised and, at the same time, disease-specific examination, is necessary, which is the intended goal in this review.

Authors' contributions

Research conception and design: Gonçalves LFG, Souto MJS, Oliveira JLM; Data acquisition: Gonçalves LFG, Souto MJS; Data analysis and interpretation: Souto MJS; Manuscript writing: Gonçalves LFG, Souto MJS, Sousa ACS, Oliveira JLM; Critical revision of the manuscript's major intellectual content: Gonçalves LFG, Sousa ACS, Oliveira JLM.

Potential Conflicts of Interest

No relevant conflicts of interest.

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Academic Association

This study is not associated to any graduate programs.

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