Value of Cardiac Magnetic Resonance Image in Brugada syndrome

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Rudy et al (**Rudic 2016**) studied with cardiac magnetic resonance imaging (CMRI) All control volunteers and BrS patients showed no contraindications for CMRI

Inclusion criteria

1. Normal physical examination, normal blood pressure (,120 and ,80 mmHg)

2. Normal ECG findings

3. No history of chest pain or dyspnea

4. No diabetes

5. Normal two-dimensional echocardiography

6. None of the controls was taking any medication.

Exclusion criteria

A. Symptoms of cardiac diseases

B. Hypertension

C. Diabetes

D. Smoking

E. *Participation in competitive or excessive sports.*

All control subjects underwent CMRI examination using the same protocol. The study was approved by the local ethics committee and written informed consent was obtained from all subjects, participating. Image acquisition and analysis. All studies were performed using a 1.5 T whole body imaging system. Images were acquired during repeated end-expiratory breath-holds. Scout images (coronal, sagittal, and axial planes) were obtained for planning of the final double-oblique long-axis and short-axis views.

To evaluate functional parameters, ECGgated cine images were then acquired using a segmented true fast imaging steady-state free-precession (FISP) sequence [echo time (TE) / repetition time (TR) 1.2/3.2 ms, temporal resolution 35 ms, in-plane spatial resolution 1.4 × 1.8 mm2 , slice thickness 6 mm, interslice gap 4 mm]. Seven to 12 short-axis views covering the whole left and right ventricle were obtained. For the evaluation of the RV anatomy and morphology, a dark blood prepared T1-weighted multi-slice turbo spin-echo pulse sequence was used to obtain axial images

from the diaphragm to the level of the RCA (i.e. to include the pulmonary outflow tract). Imaging parameters were as follows: TR ¼ 800 ms, TE ¼ 24 ms, slice thickness ¼ 4 mm, interslice gap ¼ 2 mm, and field of view ¼ 24–28 cm. In 60 of 81 patients, late gadolinium enhancement (LGE) imaging was performed. The LGE images were obtained 10–15 min after IV administration of 0.2 mmol/kg bodyweight of a gadolinium-based contrast agent (gadoterate meglumine) using an inversion recovery turbo fast low-angle shot 2D sequence or a phase-sensitive inversion recovery true FISP sequence with a slice thickness of 6 mm at the same plane position as the long- and short-axis cines in end diastole. Inversion time was adjusted individually to optimally null signal from normal myocardium and ranged typically between 250 and 300 ms. The presence of LGE was assessed visually after the consensus of two experienced readers. LV end-diastolic (ED) volumes, LV end-systolic (ES) volumes, LV stroke volume, LV ejection fraction (EF), LV cardiac output (CO), and LV myocardial mass were assessed offline from the serial short-axis true FISP cine loops using dedicated commercially available software (ARGUS, Siemens, Erlangen, Germany). In addition to volumetric measurements, one-dimensional measurements of LV ED dimensions (LVEDDs), posterior wall thickness (PWT), and maximum interventricular septum wall thickness were measured from ED short axis views. The area of the RVOT was measured on axial spin-echo images as described BY Papavassiliu (**Papavassiliu 2004**) CMRI parameters and dimensions were measured by consensus of two experienced readers, blinded to all clinical patient details.

Conclusions:

1. Mutations in the SCN5A gene lead to larger RV volume and decreased RV ejection fraction.

2. Brugada patients positive for an SCN5A mutation have a higher likelihood of diagnostic type 1 ECG than mutation negative patients. Spontaneous type 1-ECG (positive 69% versus negative mutation 38%) SCN5A mutation-positive patients have a higher likelihood of a spontaneous type 1 BrS-ECG, which is associated with a higher incidence of clinical events.

3. The presence of an SCN5A mutation was not associated with clinical events in logistic regression analysis

4. BrS patients with an SCN5A mutation reveal distinct changes in RV volumes and function when compared with those without an SCN5A mutation and healthy controls.

5. CMRI may provide additional insight to distinguish between SCN5A mutation-positive and SCN5A- mutation-negative BrS patients

Tessa et al. studied subtle abnormalities in size or function of both the RV and LV of BrS patients. They found no difference regarding both LV and RV morphology, dimensions and function in BrS patients compared to controls. LGE technique failed to show any abnormality compatible with RV myocardial fibrosis. No peculiar abnormal pattern was detected in the subgroup of patients who completely fulfilled the diagnostic criteria for BrS, in those having spontaneous type-1 ECG or in patients presenting type-1 EGC only after drug challenge. These results are in agreement with the original definition of the syndrome, that has been traditionally considered a functional electrical disorder, without any underlying structural heart disease. Martini et al (**Martini 1989**) found subtle structural cardiac abnormalities of the RV on endomyocardial biopsy and post-mortem examination in patients with BrS.

In a study of a large group of victims of SCD, a subpopulation of ARVC patients with predominant RV anterior wall involvement has been found to display an ST-segment elevation that is characteristic of BrS (**Corrado 2001**). RV fibrosis and fatty infiltration were described in two explanted hearts of BrS patients. Finally, in a study in 18 patients with a clinical phenotype of BrS and sustained VT, despite an apparently normal heart, endomiocardial biopsy revealed structural alterations in all subjects. In particular a prevalent or localized RV myocarditis was found in 14 patients, ARVC in 1 patient and cardiomyopathic changes in 3 patients. Remarkably, all the patients with cardiomyopathic changes and the patients with fibrofatty replacement were carriers of a genetic defect in the SCN5A gene, and this seems to confirm previous observation of structural changes associated with some SCN5A gene mutations. In spite of these observations, the vast majority of patients with BrS are found to possess a structural normal heart. Some imaging studies reported that a relevant percentage of BrS patients may show RV morphological and wall motion abnormalities. In particular, RV wall motion abnormalities (WMAs) were described in two electron beam computed tomography studies. Catalano et al. have shown that CMR can detect a high percentage of mild RV WMAs in BrS patients (50% vs. 17% in control subjects) These abnormalities were located mainly in the anteroapical segment, in the outflow region and in the inferior mid-ventricular segment. In this study a number of mild abnormalities of RV size and function were also detected, although only RVIT diameters enlargement retained statistical significance after correction for multiple comparisons (**Catalano 2009**).

CMR study patients presenting spontaneously type-1 ECG pattern had lower LV and RVEF, higher RV ESV, and an enlarged RVOT area compared to patients with a non-diagnostic resting ECG or controls. However, they measured RVOT area on standard axial dark-blood images, in which RVOT is sectioned with variable angles depending on the orientation of the heart in the chest and it is not possible to identify with certainty the end-diastolic phase. These results do not confirm these previous findings. Although they observed that RV WMA are detectable in a high percentage of patients with BrS, the major finding of this study is that a similar regional contractile pattern is also present in otherwise normal subjects. Data confirm that BrS patients may show some WMA of the RV but these changes are also present in healthy subjects, probably have a functional explanation, and do not indicate a diseased myocardium. The lack of LGE further indicates the absence of myocardial structural damage. These results indicate that BrS seems to occur in individuals with structurally and functionally normal heart (**Tessa 2012**).

References

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