MRI of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

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ABSTRACT

Magnetic Resonance Imaging (MRI) is currently considered as the noninvasive modality of choice for evaluation of patients with suspected Arrhythmogenic Right Ventricular Dysplasia (i.e., right ventricular dysplasia). As arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) it is included in the WHO classification of cardiomyopathies. It has the unique ability to provide tissue characterization in addition to providing functional information. This article presents the basic techniques for the evaluation of right ventricular cardiomyopathy.

Key Words: Arrhythmogenic right ventricular cardiomyopathy/dysplasia; Magnetic resonance imaging; Cardiovascular magnetic resonance; Fibro-fatty infiltration.

INTRODUCTION

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a heritable cardiomyopathy characterized by fibro-fatty infiltration of the right ventricular (RV) myocardium (Richardson et al., 1996). Affected individuals have increased incidence of sudden death due to malignant ventricular arrhythmias. The disease is more common in men and is a substantial cause of sudden deaths in athletes aged <35 yrs (Marcus et al., 1982). Fibro-fatty infiltration of the RV leads to progressive RV failure, and in the late stages of the disease process the left ventricle may also be involved (Marcus et al., 1982; Thiene et al., 1988). The diagnosis of ARVC/D is based on a set of major and minor criteria encompassing structural,
electrocardiographic, and histological criteria proposed by the Task Force of the Working Group on Cardiomyopathies in 1994 (McKenna et al., 1994). As such, accurate evaluation of right ventricular anatomy and function is crucial to diagnosis of this disease.

Conventional, noninvasive imaging modalities such as the echocardiogram and radionuclide ventriculography have several limitations, especially in patients with abnormal RV geometry. Cardiovascular magnetic resonance (CMR) imaging provides excellent anatomical details of the RV with high spatial and temporal resolution. Also, it alone has the ability to depict tissue characterization noninvasively. For the above reasons MR imaging is considered the noninvasive imaging modality of choice to evaluate ARVC/D (Pennell and Casolo, 1997). The improved contrast between the blood pool and the myocardium allows accurate and highly reproducible quantitation to be performed, making it ideal for follow-up evaluation and to study the disease progression.

**MR Imaging of Right Ventricular Cardiomyopathy**

The MR imaging protocol in right ventricular cardiomyopathy is aimed at recognizing two important aspects of the disease process: 1) global and regional right ventricular morphology, 2) global and regional right ventricular dysfunction, and 3) fibrofatty infiltration of the right ventricle. Hence the protocol includes 1) bright-blood cine imaging to visualize right ventricular global and regional function, and 2) black-blood imaging to identify intramyocardial fatty infiltration (Bluemke et al., 2003; Castillo et al., in press). The optimal protocol for ARVC/D is directly proportional to the scan time (duration of the scan), and in our experience it is around 45 minutes to 1 hour. The equipment parameters required to run the protocol are shown in Table 1.

For bright-blood imaging, steady-state free precession imaging is the preferred technique (FIESTA, true FISP, Balanced Fast Field Echo). If those cine sequences are not available, segmented k-space gradient echo images [e.g., fast low angle shot (FLASH); fast cardiac gated gradient echo (FASTCARD)] can be used. For black-blood techniques, breath-hold imaging with double inversion recovery fast spin echo (FSE) techniques are preferred to traditional spin echo (SE) imaging. These techniques substantially shorten the imaging time and are devoid of respiratory motion artifacts. Black-blood inversion prepared, half-Fourier single-shot turbo spin echo (HASTE) imaging has not been systematically evaluated, but is currently not recommended due to blurring of detail with this sequence. All images are optimally performed during end-expiratory breath-hold.

**MR Imaging Protocol**

**Sequence 1: Sagittal scout: Any rapid image localizer**

**Sequence 2: Axial black-blood images**

Axial imaging plane provides the best view of the right ventricular anterior wall and to the proximal parts of the right ventricular outflow tract. This imaging plane is useful to demonstrate intramyocardial hyperintense T1 signals and to evaluate for outflow tract enlargement. Prescribe the axial images starting from the diaphragm to the pulmonary artery (Fig. 1). Black-blood images are acquired at the authors’ institutions using double inversion recovery, fast spin echo sequence with blood suppression. If the center has a dedicated cardiac coil, best results are obtained using the anterior coil elements only (posterior coil switched off) to prevent a “wrap around” artifact. Also, the authors recommend using an anterior saturation band (Fig. 1). The scan parameters for the sequence are as follows: TR=2R-R intervals, TE=5 msec (minimum full), slice thickness=5 mm, interslice gap = 0–5 mm, and field of view (FOV)=24–32 cm.

### Table 1. Equipment parameters for basic protocol.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Specification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coil type</td>
<td>Cardiac phased array coil (or Torso phased array coil)</td>
</tr>
<tr>
<td>Gradient coil strength</td>
<td>Minimum 20 mT/m</td>
</tr>
<tr>
<td>Cardiac gating</td>
<td>Yes, preferably fiber optic cables</td>
</tr>
<tr>
<td>Breath-hold imaging</td>
<td>Yes</td>
</tr>
<tr>
<td>Respiratory gating</td>
<td>Yes, if breath-hold imaging not possible</td>
</tr>
</tbody>
</table>

Note: If the patient is known to have frequent ventricular ectopy, the authors recommend the use of oral Metoprolol 50 mg, at least 1 hour prior to the procedure provided that the patient has no contraindications. If ventricular arrhythmias are frequent and will substantially impact image quality, the exam should be terminated at this point.
Sequence 3: Sagittal black-blood images
Since the right ventricular outflow tract bends backwards towards the pulmonary valve, axial slices may not suffice, in which case two additional sagittal planes are required. This is especially important when there is suspected fatty infiltration in these more distal parts of the right ventricular outflow tract (RVOT). The same image parameters as used for Sequence 2 can be applied. Inclusion of the subvalvular parts of the RVOT has to be ensured.

Sequence 4: Axial bright-blood cine images
Cine imaging in the axial plane is optimal to assess right ventricular global and regional function visually. Also, one can assess the right ventricular and atrial chamber enlargement in this plane. As previously mentioned, the most preferred method of cine imaging is by using the steady-state free precession technique. This allows better endocardial definition when compared to fast gradient echo (FGRE). For best results while using steady-state free precession, one should manually inspect resonance peaks to ensure that the center frequency is water.

Figure 1. Sagittal scout image showing the prescription of axial cine sequence. Note that the coverage for the axial images starts from the diaphragm to the pulmonary artery. Also shown in the figure is the placement of anterior saturation band.

Figure 2. Prescription of long-axis scout cine image on axial image. The localizer is placed in the long axis of the left ventricle parallel to the interventricular septum. The grid line is drawn from the left ventricular apex to the mid-mitral valve plane, and parallel to the interventricular septum.
Use the same or at least a very similar prescription for the axial cine images as in the previous series, i.e., starting from the diaphragm to the pulmonary artery. Scan parameters for the bright-blood cine images as follows: TR=3.5, TE=1.2 msec, flip angle=45°, slice thickness=5–8 mm, inter-slice gap=0–5 mm, and FOV=24–36 cm, 20 views per segment. The flip angle may vary depending on the manufacturer.

**Sequence 5: Sagittal bright-blood cine images**

Using the same orientation as for Sequence 3, the more distal parts of the right ventricular outflow tract should be assessed using the same bright-blood technique as in Sequence 4.

**Sequence 6. Vertical long-axis scout images**

On a mid-cavity, axial image, prescribe a long-axis scout image along a line drawn from the left ventricular apex through the mid-mitral valve plane, drawn like a bisecting line through the left ventricle as shown in Fig. 2. Prescribe one slice, any rapid scout sequence.

**Sequence 7: Horizontal long-axis scout**

On the long-axis scout image, prescribe a four-chamber scout cine image as shown in Fig. 3. This view provides a horizontal, long-axis view of the...
heart, which does not equal the true four-chamber view, but allows the definition of the short-axis view. The scanning parameters are identical to those of Sequence 5.

Sequence 8: Short-axis cine
This sequence is prescribed from the four-chamber view. It is helpful to prescribe off an end-diastolic, four-chamber image. Coverage for this sequence should start from above the mitral valve plane to the apex of the heart as shown in Fig. 4. This is important as this view is often used for quantification of ventricular volumes. Quantification of ventricular volumes is performed by manually contouring the end-diastolic and end-systolic frames of the entire RV, using a summation of disks method (Simpson’s Rule), with integration over the image slices. The parameters for this sequence are identical to those of Sequence 5.

Sequence 9: Short-axis black-blood images
This view is useful to assess the inferior (diaphragmatic) border of the right ventricle. Care should be taken to adjust the field of view appropriately to minimize ‘‘wrap artifact.’’ In scanners with surface coil, we recommend using anterior coil elements only and switching off the posterior coil to reduce the wrap artifact. The parameters for this sequence are the same as those for Sequence 2.

Sequence 10: Four-chamber view cine
In the short-axis images, the true, four-chamber view similar to the echocardiographic four-
chamber view can be obtained by defining a line from the center of the left ventricle to the widest part of the right ventricle (Fig. 5). In this view one can assess the relative chamber sizes of the left and right ventricles and ventricular function visually.

**Sequence 11 (Optional Sequence): Axial black-blood images with fat suppression**

This sequence is optional and should be applied if there is evidence of fatty infiltration in one of the sequences 2, 3, or 9. It usually adds 10 minutes to the total scanning time. Repeat series 2, 3, and 9 with chemical selective fat suppression.

**ANTICIPATED RESULTS**

The goal of MR imaging in ARVC/D is to accurately depict RV structure and function. When evaluating RV structure special attention should be paid to the infero-basal part of the RV inflow, RV apex, and the anterior wall of the outflow tract, as these are the commonly involved sites in ARVC/D. Fat infiltration can often be difficult to visualize, as epicardial fat is normally present abundantly near the atrio-ventricular groove and near the RV apex. Fig. 6 shows an axial, black-blood image from a normal volunteer demonstrating the normal distribution of epicardial fat in the RV. A clear line of demarcation exists between the epicardial fat and the RV myocardium in normal subjects, and a disruption of this line is often seen in patients with ARVC/D (Fig. 7). Also, there is increased trabeculation of the RV with prominent moderator band in patients with ARVC/D. Assessing RV function visually can be very challenging as the RV has a complex morphology and the contraction pattern differs from that of the left ventricle. We recommend assessing RV function from the axial, bright-blood cine images. The RV on axial images starts off as a large triangle in diastole, which becomes a smaller triangle in systole. Most of the contraction occurs in the long axis of the RV from the movement of the tricuspid valve towards RV apex. The RV anterior wall should be examined for bulging and aneurysms (Fig. 8), as these are very specific morphological abnormalities associated with ARVC/D. In our experience we find that global, functional abnormalities of the RV are present in a majority of ARVD/C patients, and are more reliable for the diagnosis. The definition of RV endocardial borders may be difficult in the valvular areas, and results may be limited by a certain interstudy and interobserver variability. Nevertheless, quantification of RV volumes and ejection fraction are recommended, as these are more reproducible than qualitative estimates.

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**REFERENCES**


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