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MR and CT imaging of Arrhythmogenic Cardiomyopathy

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Right ventricular structure and functional alterations are important criteria for the diagnosis of arrhythmogenic cardiomyopathy (AC). As such, accurate and reliable evaluation of RV is desirable both for establishing the diagnosis and for follow up evaluation of AC. Right ventricle has a unique three dimensional geometry and complex contraction pattern¹. Thus conventional non invasive imaging modalities like the echocardiography and radio nuclide ventriculography are limited in their assessment. Recently Magnetic resonance (MR) imaging and computed tomography (CT) have emerged as robust tools to evaluate the RV in patients with suspected AC^{2–6}. The noninvasive nature of these investigations, multiplanar capability, and unique ability to provide tissue characterization are ideal for assessment of AC. Both modalities have the ability to provide direct evidence of fatty infiltration and structural alterations of the RV. The purpose of the current chapter is to discuss the current status, strengths and limitations of MR imaging and cardiac CT, in evaluation of AC.

MR Imaging in Arrhythmogenic Cardiomyopathy

Over the last decade there have been significant improvements in MR imaging hardware such as dedicated cardiac coils, faster imaging, and excellent blood pool contrast with steady state free precession imaging, which has made MR imaging the imaging modality of choice for cardiac applications. ECG gating and breath-hold imaging have reduced motion artifacts and improved tissue contrast is achieved by inversion recovery black-blood imaging techniques⁷. Steady state free precession imaging (SSFP) pulse sequence, have resulted in better delineation of endocardial borders, enabling accurate and reproducible volumetric measurements⁸. MR imaging is now routinely used for quantitative and qualitative assessment of cardiac function. Among the current cardiac MR imaging applications in cardiomyopathies, the greatest potential as well as biggest challenges are in the diagnosis of AC. Routinely used imaging planes are sub-optimal for RV evaluation and the technique of AC imaging involves unconventional imaging planes. Furthermore the lack of familiarity of the MR readers with the RV contraction pattern and the normal epicardial fat distribution poses challenges for accurate and reproducible reporting.

Despite the above limitations, MR imaging is uniquely suited for evaluation of AC. It has the ability to non-invasively provide tissue characterization for detection of fat infiltration and fibrosis in the RV, which are the histopathologic hall marks of AC^{9–11}. Quantitative and reproducible data on RV volumes and function are not only useful in establishing the diagnosis, but also for follow-up of AC patients. Finally, the multiplanar depiction of RV

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wall motion is invaluable in assessing RV regional contraction abnormalities, which are often the earliest manifestation of AC.

MR imaging protocol

The MR imaging protocol in right ventricular dysplasia is aimed at recognizing two important aspects of the disease process: (a) fibro-fatty infiltration of the right ventricle, (b) global and regional right ventricular dysfunction. Hence the protocol includes (a) black-blood imaging to identify intramyocardial fatty infiltration, (b) bright-blood cine imaging to visualize right ventricular global and regional dysfunction and (c) myocardial delayed enhancement images. Table 1 shows the MR imaging protocol used at our center. Typically axial and short axis black blood images are acquired for detecting fat infiltration. Axial, short axis and four chamber images are acquired for assessing RV and left ventricle (LV) wall motion. Delayed enhanced images are obtained in the same views as the functional images. 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. For bright-blood imaging steady state free precession imaging is the most preferred technique (FIESTA, true FISP, Balanced Fast Field Echo). If those cine sequences are not available, segmented k-space cine gradient echo images (e.g.; fast low angle shot—FLASH; fast cardiac gated gradient echo—FASTCARD) can be used. All images are optimally performed during end expiratory breath-hold. Delayed enhanced images are obtained at our center using 2D inversion recovery prepped gradient echo imaging or Phase Selective Inversion Recovery (PSIR) sequences. It is important to use a dedicated cardiac coil for imaging and to use the anterior coil alone while obtaining the black blood images. The latter prevents wrap around artifacts. Also not infrequently these patients present with frequent ventricular premature beats that cause blurring of the image due to inadequate gating. If the patient is known to have frequent ventricular ectopy, we 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 then the resulting scan quality will be poor and the test becomes uninterpretable.

MR imaging findings in Arrhythmogenic Cardiomyopathy

Broadly, MR imaging abnormalities in AC can be grouped into two major categories: 1) morphological abnormalities, and 2) functional abnormalities. Morphologic abnormalities include intramyocardial fat deposits, focal fibrosis, wall thinning, wall hypertrophy, trabecular disarray, and RV outflow tract enlargement. Functional abnormalities include regional contraction abnormalities, aneurysms, RV global dilation /dysfunction, and RV diastolic dysfunction. The sites of involvement of these abnormalities are observed in the “triangle of dysplasia”, which is constituted by the inferior-sub tricuspid area, RV apex and the RV infundibulum (Figure 2). The goal of MR imaging in AC is to accurately assess the RV for the presence or absence of these abnormalities, to aid in the diagnosis and follow-up of patients.

Intramyocardial fat

Abundant epicardial and intrapericardial fat is commonly found over the RV but intramyocardial fat infiltration in the right ventricle is rare. A clear line of demarcation exists between the epicardial fat and the RV myocardium in normal subjects Figure 3a & b) and a disruption of this line is often seen in patients with AC. Shown in Figure 4 is a classic

example of fat infiltration in AC. Note the finger like projections of the epicardial fat (4a) causing a signal void in the myocardium in the fat-suppressed images (figure 4b).

The prevalence of intramyocardial fat in AC on T1 weighted SE imaging has ranged from 22% to 100% in different studies^{2, 10, 12–14}. We used breath-hold double inversion recovery fast spin echo technique to evaluate intramyocardial fat in AC, and found a high intramyocardial T1 signal (fat) in 9 of 12 patients (75%) who were prospectively diagnosed using the current Task Force criteria¹⁰. The differences in incidence of fat signal among various studies in AC are largely based on differences in patient selection, different imaging techniques used and most importantly on the definition of abnormal intramyocardial fat. It is known from the autopsy studies that the degree of fat infiltration is associated with the stage of the disease. Advanced AC is often associated with greater degree of fat infiltration and there may be no evidence of macroscopic fat infiltration on imaging in the early stages of the disease¹⁵. Current MR imaging resolution however is insufficient to diagnose early stages of the disease, even when there is clear evidence of fat infiltration on histopathology.

Fat infiltration when present is predominantly seen in the RV outflow tract and the anterior RV free wall and the basal RV especially close to the RV inlet. Shown in Figure 4 is the classic appearance of fat infiltration in AC. Note the disruption of the border between the epicardial fat and the RV wall. Fat suppressed image shows multiple signal voids within the RV myocardium. Detection of fat infiltration close to the apex however is challenging as this region is usually very thin (2mm–4mm) and there is abundant fat close to the cardiac apex. In any case, the use of spectrally selective fat suppression with the black blood sequences provides additional evidence of fat infiltration due to high contrast between epicardial fat and the RV myocardium. The accurate delineation of the underlying RV free wall increases the confidence of diagnosis for the MR physician.

Although detection of fat infiltration on MR imaging is moderately sensitive to the disease process, the specificity of this finding is unclear. Detection of intramyocardial fat on MR imaging requires considerable experience and over reliance on this finding alone often leads to mis-diagnosis of AC. Excessive epicardial fat can often masquerade as fat replacement of the RV (Figure 5a & b)). In some cases this will also distort RV regional wall motion further confusing the issue. Steroid use, alcoholism, obesity and old age have all been associated with fat infiltration^{16–18}. Nevertheless the classic appearance of segmental fat infiltration appears to be unique to AC. However, it cannot be underscored that reliable interpretation of this finding requires considerable expertise, which is often difficult to establish in this rare disease.

Wall Thinning and Wall hypertrophy

RV wall thinning is defined focal abrupt reduction in wall thickness to < 2mm, surrounded by regions of normal wall thickness. This finding is uncommon in AC and is often over-called especially close to the RV apex where the RV tends to be normally very thin. Wall thinning is due to progressive loss of epicardial and myocardial layers that leaves a thin rim of endocardial cells, which are usually not involved until late in the disease process. In our series we found wall thinning in less than 25% of patients who met the Task Force criteria. In our experience wall thickness is often difficult to assess due to adjacent high epicardial fat signal and motion artifacts.

Wall hypertrophy, defined as RV wall thickness > 8mm is usually an MR appearance wherein it is impossible to differentiate the true RV wall from the overlying epicardial fat as the line of separation is completely disrupted. This finding is seldom observed in pathologic specimens as the true RV myocardium is measured exclusive of the epicardial fat¹⁹. In vivo this differentiation is sometimes not possible due to extensive fibro-fatty infiltration with

loss of distinction between epicardial fat and the true myocardium. In such cases the RV wall appears hypertrophied with MR images showing islands of grey muscle surrounded by bright signals compatible with fat. Fat suppressed image reveals multiple signal voids within the RV myocardium in locations that showed hyperintense signals in the non-fat suppressed images (Figure 4b).

Trabecular Disarray

Heavy trabeculation of the RV is frequently observed in AC, more so in patients with RV dysfunction. Hypertrophy of the trabeculae and the RV endocardium which compensate for the epicardial myocyte loss is thought to result in this appearance. Occasionally giant Y shaped trabeculae and hypertrophy of the moderator band are observed in patients with AC. This finding has been equated to the angiographic finding of deep fissures with a 'pile d'assiettes' (stack of plates) appearance. RV trabecular disarray is a finding that is not specific for AC and may be present in any condition that results in RV hypertrophy or enlargement.

RV Outflow Tract Enlargement

In our experience the RV outflow tract (RVOT) is the most common location for localized AC. The right ventricular outflow is usually equal to or marginally smaller than the aortic outflow tract at the level of the aortic valve. An exception to this rule is pediatric patients in whom the RVOT may be larger than the left ventricular outflow. Presence of an enlarged RVOT beyond adolescence is uncommon. More important than a simple enlargement is a dysmorphic appearance of the outflow tract (Figure 6). Abnormal appearance of the RVOT, which is dyskinetic in systole, is highly suggestive of AC in the absence of pulmonary hypertension.

MR imaging fibrosis in Arrhythmogenic Cardiomyopathy

One of the pathologic hallmarks of AC is fibrosis of the RV that accompanies fatty infiltration. Myocardial delayed enhancement MR imaging allows for non-invasive detection of fibrosis in the RV. Delayed enhancement detected by MR imaging shows excellent correlation with histopathology and an inverse correlation with global RV function²⁰. Further, areas of delayed enhancement often show wall motion abnormalities on cine imaging. Common sites for delayed enhancement are the basal sub-tricuspid region, the acute angle of the right ventricle and the RV outflow tract (Figure 7). Presence of delayed enhancement also predicts inducibility of re-entrant ventricular tachycardia in AC. One of the most important uses of delayed enhancement imaging is its role in differential diagnosis. Presence extensive bi-ventricular delayed enhancement in a patient being evaluated for AC should suggest alternative diagnoses such as sarcoidosis or myocarditis. The myocardium of the interventricular septum is rarely involved in AC and frequently involved in sarcoidosis (Figure 8). The differentiation of isolated RV myocarditis and sarcoidosis involving the RV from AC is often difficult using MR imaging alone. Both sarcoidosis and myocarditis can show patchy involvement of the LV and fat infiltration is strikingly absent. Clinical findings such as conduction disease, multisystem involvement and hilar lymphadenopathy suggest sarcoidosis even when the MR imaging findings are indistinguishable.

Left Ventricular Involvement in Arrhythmogenic Cardiomyopathy

Desmosomal mutations affect both ventricles; hence it has been argued that AC is a biventricular disease with predominant involvement of the right ventricle. Studies to date suggest that LV involvement may be dependent on the underlying mutation. Mutations in Desmoplakin and Plakoglobin have been associated with bi-ventricular AC or in some cases

left dominant AC²¹. Mutations in plakophilin-2 (PKP2) on the other hand result in the classic form of AC with minimal LV involvement, at least minimal global functional involvement²². Histologic studies however have reported LV involvement in up to 70% of patients, but these are typically autopsy data and the extent of disease is unknown.

Left ventricular MR imaging findings in AC are unique and are mostly localized to the basal inferior and infero-lateral left ventricle (Figure 9). This is typically in the form of fat infiltration that extends from the epicardium in to the myocardium with sparing of the endocardium. Fat infiltration in this location is not seen in any other disease, thus making this a finding that is unique to AC. Even with extensive amount of fat, global LV function is not affected possibly because this is localized and well compensated by the remainder of the LV. The same site may show delayed enhancement, however, this may be difficult to reconcile from fat infiltration unless a fat suppressed delayed enhanced sequence is performed. Severe LV dysfunction and LV arrhythmias are rare in PKP2 related AC.

MR Assessment of Cardiac Function in Arrhythmogenic Cardiomyopathy

The techniques to image cardiac function are called “bright-blood” techniques, derived from the appearance of intracavitary blood. Consecutive images that are acquired with a high temporal resolution can be viewed dynamically to generate functional information. Although a number of sequences exist for bright-blood imaging, steady state free precession imaging is the most preferred technique (FIESTA, true FISP, Balanced Fast Field Echo). SSFP sequences result in improved contrast between the blood pool and the myocardium compared to segmented *k*-space cine gradient echo images. If SSFP is not available, segmented *k*-space cine gradient echo images (e.g.; fast low angle shot—FLASH; fast cardiac gated gradient echo—FASTCARD) can be used. Conventional gradient recalled (GRE) imaging sequences rely on flowing blood to generate bright blood. In the dysfunctional RV, blood velocities are reduced and signal intensity decreases with conventional GRE imaging. With SSFP the signal intensity remains high since the signal intensity is proportional to T2 time.

Ventricular volumes and mass using bright-blood imaging have been shown to be accurate and reproducible and MR imaging is considered the standard of reference^{23, 24}. Thus MR imaging is ideal for non invasive assessment global RV functional in patients with suspected AC and their first degree relatives.

Functional Abnormalities in Arrhythmogenic Cardiomyopathy

Global RV dilation/dysfunction

Fibro-fatty replacement of the RV in AC eventually leads to RV dilation and dysfunction. Moderate to severe RV dysfunction constitutes one of the major task force criteria for AC. Recent refinements to the task force criteria have resulted in introduction of quantitative cut-offs for global RV volumes and function for the diagnosis of AC^{25, 26}. RV dysfunction is often present in the majority patients who meet the task force criteria especially in the absence of a family history of AC. Also there is a linear correlation between the RV end diastolic volumes and the duration of symptoms suggesting the progressive nature of the disease. However, asymptomatic first degree relatives who are diagnosed solely by virtue of their family history or due to presence of a pathogenic mutation often tend to have preserved RV function. Serial quantitative volumetric assessment of RV may play an important in assessing RV function in first-degree relatives.

Regional Dysfunction

MR imaging has replaced RV angiography in RV regional function assessment as the former technique is non-invasive and is devoid of ionizing radiation. Regional RV dysfunction is generally thought to be due to focal fibro-fatty infiltration, which precedes changes in global ventricular function. As such, regional dysfunction is seen in the same areas as the fat infiltration; the RVOT, the anterior RV and the basal RV inlet. Shown in Figure 10 is a basal aneurysm in a patient with advanced AC. Regional functional abnormalities of the RV described in AC include focal hypokinesis (wall thickening of < 40%), akinesis (systolic wall thickening of < 10%), dyskinesis (myocardial segment, which moves outward in systole) and aneurysms (segments with persistent bulging in diastole, and dyskinetic in systole). Studies have consistently reported high incidence of regional dysfunction in AC. The areas of dysfunction corresponded to the areas of signal abnormality observed on black blood MR imaging. The presence of signal abnormality associated with abnormal wall motion is more suggestive of AC compared to either of them alone. In our series, 67 % of the patients had regional contraction abnormalities, which correlated to the area of adipose replacement on MR imaging¹⁰. RV aneurysmal dilation is often seen in advanced AC and coexists with global RV dysfunction. Care should be taken while interpreting RV regional function from axial views, as the axial plane is not orthogonal to the inherent axis of the heart and systolic bulging can be seen even in normal volunteers. Correlation with horizontal long axis view may be helpful to avoid misinterpretation of RV regional function.

Genotype-Phenotype correlation

Genotype-phenotype correlations suggest that PKP2 mutation usually present with the classic, right-dominant disease²⁷, while other series with a relatively higher prevalence of desmoplakin mutations consist of patients who show a more diverse phenotype, including the so-called left dominant AC²¹. Further, the presence of the mutation is associated with higher prevalence of LV fat infiltration compared with AC patients in whom no mutation is found. In mutation positive first degree relatives we have observed minor contraction abnormalities limited to the RV outflow tract and the RV basal region²⁷. These regional contraction abnormalities are best described as a focal crinkle in the RV during systole resembling an “Accordion”(Figure 11). Although these findings may be sensitive, they come at a price of reduced specificity and poor reproducibility limiting their use in the diagnosis of AC. Finally, preliminary genotype-phenotype data suggest that disease severity is greater in double mutations carriers, further emphasising the need to screen all known disease-causing genes even after isolation of a pathogenic mutation.

Role of MR Imaging in Diagnosis of Arrhythmogenic Cardiomyopathy

Diagnosis of AC is challenging especially in early stages of the disease. The lack of diagnostic gold standard makes it difficult to define the sensitivity and specificity any single modality in diagnosing AC. Benign arrhythmias arising from the RV outflow tract are frequently encountered in clinical practice and the presence of characteristic structural abnormalities differentiates AC from the benign idiopathic ventricular tachycardia²⁸. As such the role of MR imaging in accurate phenotyping is invaluable. However, qualitative findings such as intramyocardial fat infiltration and subtle wall motion abnormalities need considerable expertise for interpretation. Isolated areas of fat replacement are not specific to AC and have been reported in elderly patients, patients receiving long-term steroids, and in other cardiomyopathies. Recently we have observed marked lipomatous infiltration of the right ventricle in young non-obese individuals who were evaluated for non-sustained ventricular arrhythmias¹⁸(Figure 8). None of these patients had global or regional functional abnormalities and thus appeared to be a distinct group of patients defined by MR imaging that should be differentiated from patients with AC. Over-reliance on the presence

of intramyocardial fat has in fact resulted in a high frequency of "misdiagnosis" of AC²⁹. In our experience this finding alone is neither sensitive nor specific for the diagnosis. Our experience with MR imaging of autopsy hearts lead us to conclude that the achievable spatial resolution in current state-of-the-art clinical protocols substantially limits the capability to detect subtle RV intramyocardial fatty changes. Since the disease is so rare, most imaging centers have little or no experience with diagnosis of AC. Technical problems in imaging patients with arrhythmias, lack of a standardized protocol for AC, and lack of experience by imaging physicians suggest that MRI should be only one part of a comprehensive evaluation for these patients. According to the task force criteria, MR imaging provides information related to RV size, global and regional function and aneurysm formation. Quantitative cut-offs are now a part of the task force criteria and relying on quantitative evaluation of RV size and function is advisable and avoids the risk of over diagnosis of AC.

One of the most important roles of MR imaging is to reliably identify patients who require additional invasive testing. A completely normal MR study in a patient with no abnormalities on electrocardiography or echocardiography is reassuring, and such patients may not need invasive testing (angiography/biopsy) in the absence of other clinical criteria. If signal abnormalities and wall motion abnormalities coexist, invasive testing should be undertaken to confirm the findings. Minor structural abnormalities i.e. signal abnormalities in the absence of wall motion changes presents a challenge as further evaluation of such patients is unclear. Adherence to Task Force criteria is recommended, and these "minor" criteria may not necessitate invasive testing. Further, it must be recognized that AC Task Force criteria do not currently recognize fat signal on MR (or CT) as a diagnostic criterion for the disease.

Finally, when evaluating RV structure special attention should be paid to the RV inflow, RV apex and the outflow tract, as these are the commonly involved sites in AC. 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, as these are morphologic abnormalities associated with AC. Quantification of RV volumes and ejection fraction should be performed routinely, as these are more reproducible than qualitative estimates and can easily be applied using the current criteria for the purposes of diagnosis.

Computed Tomography of Arrhythmogenic Cardiomyopathy

Following a similar trajectory to MR imaging, there has been a continuous and accelerated development of CT technology, particularly in the last 10 years focused towards cardiac imaging. Currently available multidetector CT (MDCT) imaging provide excellent spatial resolution, and allow accurate high-resolution assessment of morphological detail of both the cardiac chambers^{6, 30, 31}. The use of a non-ionic contrast agent provides excellent contrast resolution with clear delineation of the ventricular endocardium. Multiple cardiac phases can be extracted, with animated movies of the beating heart made available for visual assessment of global and regional function^{32, 33}. Quantitative determinations of ventricular mass, right and left ventricular volumes, and global ventricular function can be performed in a variety of cardiac pathologic states.

One of the main advantages of CT over MR imaging is that it is fast, easy to perform, and has more reliable image quality. Although images are acquired only in the axial plane, the acquisition of a 3-dimensional dataset allows reformatting in any desirable plane. For the

above reasons CT is a clinically valuable, non-invasive tool for assessment of myocardial pathology.

Computed Tomography imaging has been used to evaluate AC and has the ability to depict the characteristic abnormalities in AC such as abnormal RV morphology and fat infiltration. The ability of conventional CT to detect intramyocardial fat in AC was first reported by Villa et.al,³⁴ in a series of seven patients with AC, and subsequently Sotozono et al³⁵ provided biopsy confirmation of CT findings. Other investigators have shown the utility of CT in AC including the assessment of LV abnormalities in particular LV fat infiltration. Intramyocardial fat appears as a hypointense signal on CT and this is particularly prominent in the LV wall (Figure 12). Intramyocardial fat is defined based on tissue attenuation values. The attenuation value for epicardial adipose tissue is around -65 ± 10 Hounsfield units (HU), and 5 to -17 HU for intramyocardial fat which is far less than that of myocardium. Using the above values Tada et al³⁶ showed that this was specific for AC as none of the control subjects, and no patient without AC showed any evidence of intramyocardial fat, or any other qualitative features of AC. Further, there is a good correlation between areas of fat detected on CT and abnormal RV endocardial voltage on electroanatomic mapping.

Hamada et al.³⁷ imaged four AC patients with RV arrhythmias who had abnormalities on electrocardiography and angiography using EBCT. Quantification of ventricular volumes was performed on cine mode scanning, which showed regional dysfunction and depressed global RV function respectively. Recently a case of AC was evaluated with 64 detector CT and with contrast enhanced volume mode scanning they were able to demonstrate morphologic abnormalities consistent with AC such as RV enlargement, excessive trabeculations, fatty infiltration, and marked RV hypokinesis³⁸.

Volumetric assessment of the RV is sub-optimal using CT imaging. Problems that we faced using MDCT include artifacts due to ICD leads, mis-registration of slices, problems with reformatting the short axis data from axial images, all of which undermine the accuracy and reproducibility of the data. Most of the patients who were evaluated using CT imaging at our center had implantable defibrillator hardware which precluded MR imaging. Nevertheless, all the morphologic features are well depicted by CT and chamber dimensions can easily be obtained using the current generation CT imaging.

Role of CT in AC

Most of centers including ours currently rely on MR imaging instead of CT imaging for evaluating patients with suspected AC mainly because the former is devoid of radiation. The temporal resolution of CT imaging is sub-optimal to allow accurate quantification of volumes. However, most patients who are diagnosed with AC receive implantable defibrillators for prevention of sudden death. In this cohort CT remains the only imaging technique to assess RV structure and function for serial morphologic evaluation. An additional use of CT imaging is to assess the mediastinum for presence of lymphadenopathy, a marker of sarcoidosis, which occasionally mimics AC³⁹. CT imaging also has the ability to assess the lung fields for nodules or granulomas suggestive of sarcoid involvement of the lung. CT is also useful in the occasional patient who has frequent premature beats resulting in arrhythmic artifacts on MR imaging, and also in patients who are claustrophobic.

MDCT radiation can be quite high, exceeding conventional angiography by a factor of 2 when performing retrospective gating⁴⁰. Thus, MDCT may not be optional for “screening of AC in first degree relatives. Despite the above limitations, CT does provide certain advantages over MR imaging in terms of consistency in image quality, scan time, operator dependency etc. With increase in familiarity of radiologists with the use of helical CT for

AC and with the advances in both the temporal and spatial resolution, CT imaging may play an important role both in the diagnosis and in the follow-up of patients with AC.

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Figure 1.

Figure 2.

Figure 3.

Figure 4.

Figure 5.

Figure 6.

Figure 7.

Figure 8.

Figure 9.

Figure 10.

Figure 11.

Figure 12.

Table 1

MR Imaging Protocol For ARVD

Imaging Sequence	Imaging Plane	Remarks
Black Blood Images	Axial	Assess for RV and LV fat infiltration
	Short axis	
Bright Blood Images	Axial	Assess for RV regional function
	Four Chamber	RV size and LV function estimation
	Short axis	Optimal for quantification of RV volumes
Myocardial Delayed Enhancement	Axial	Assess for fibrosis
	Four Chamber	
	Short axis	