

Role of cardiac imaging evaluation of patients with documented or suspected ventricular arrhythmias

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Sudden cardiac death (SCD) accounts for over 400,000 deaths annually in the United States alone.¹ The majority of these are due to malignant ventricular arrhythmias.² Coronary artery disease (CAD) is the most common underlying heart disease. Inherited heart disease such as hypertrophic cardiomyopathy, Long QT syndrome, and arrhythmogenic right ventricular dysplasia (ARVD) account for less than 5% of the deaths.³⁻⁵ Patients who present with ventricular arrhythmia and who have structural heart disease are at high risk for SCD events.⁶⁻⁸ Ascertaining the etiology of the underlying heart disease is important for management of the patient and for prognostic reasons. Recent advances in cardiac imaging have substantially improved our ability to diagnose the etiology of ventricular tachycardia (VT), assess the extent of the disease, and plan management strategies. This review summarizes our current understanding of the role of cardiac imaging in the diagnosis and management of patients with ventricular arrhythmias.

ROLE OF IMAGING IN DIAGNOSIS

Ventricular arrhythmias occur both in structurally normal hearts and in disease states.^{9,10} The overall prognosis of ventricular arrhythmias occurring in structurally normal hearts is excellent.¹¹⁻¹³ On the other hand, the prognosis in patients with structural heart disease is determined by the nature of heart disease and the severity of ventricular arrhythmias. Common imaging modalities used to evaluate patients with ventricular arrhythmias include 2D echocardiography, computed tomography (CT), and magnetic resonance (MR) imaging. Left ventricular ejection fraction is one of the most

useful predictor of future arrhythmic events. Although some form of assessment of ventricular function is desirable, this, however, does not provide additional information over above identifying high-risk individuals. Over the last decade, advances in MR imaging and CT imaging have led to better understanding of the substrate and hence improved our ability both for diagnostic and therapeutic interventions.

MR Imaging for Diagnosis

MR imaging has evolved to be extremely useful in the diagnosis of patients with suspected arrhythmias, more importantly in patients with inherited arrhythmic disorders.

It has the ability to provide multiplanar imaging noninvasively, which has led to a tremendous increase in its utilization.^{14,15} The ability to provide tissue characterization in addition to functional information has made it the imaging modality of choice in patients with ventricular arrhythmias of right ventricular (RV) origin.¹⁶⁻¹⁸ Depiction of the location and extent of myocardial fibrosis provide clues to the underlying etiology of cardiomyopathy and the site of origin of ventricular arrhythmias.¹⁹ The latter is often useful to guide curative therapies such as radiofrequency ablation.

MR Imaging in Right Ventricular Arrhythmias

Arrhythmias arising from the right ventricle often have a left bundle branch block (LBBB) morphology on the surface ECG. Differential diagnosis includes: (1) Idiopathic right ventricular outflow tract tachycardia, commonly referred to as idiopathic VT, (2) ARVD, (3) sarcoidosis, and (4) myocarditis.

Differentiating these conditions is very important as idiopathic VT is a benign condition with a favorable prognosis, compared with ARVD,²⁰ sarcoidosis, or myocarditis. Diagnosis of idiopathic VT is often based on the morphology of ventricular arrhythmia that is consistent with the RV outflow origin of the VT (LBBB inferior axis) and the lack of structural heart disease on imaging.

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On the other hand, ARVD is a genetic cardiomyopathy with an autosomal dominant inheritance pattern. It carries a significant risk of SCD due to malignant ventricular arrhythmias requiring implantable defibrillator implantation. Once the diagnosis of ARVD is entertained, it is mandatory to screen the first-degree relatives of the patient for evidence of ARVD. Sarcoidosis can often respond to steroid therapy and hence accurate diagnosis is crucial.

Arrhythmogenic right ventricular dysplasia. ARVD is a heritable cardiomyopathy characterized by the fibrofatty replacement of RV myocardium leading to RV failure and arrhythmias.²¹ The exact prevalence is unknown, but it is estimated to be 1:5000 in the United States. It often affects young men who have an athletic life style. Palpitations and exertional syncope are the most common presenting symptom. Arrhythmias in ARVD have a LBBB morphology, and the axis of the VT provides clues to the site of origin of the VT. The disease often affects the RV outflow tract, the base of the RV, and the RV apex, collectively termed as the triangle of dysplasia.²²

MR imaging is uniquely suited to evaluate this condition as it not only provides excellent functional information for the RV, but in addition can depict fibrosis and fatty infiltration in the RV.^{18,19} Figure 1 shows the common MR imaging findings in ARVD. These include RV dilation and dysfunction, RV regional hypokinesis, and focal aneurysmal dilation of the RV especially in the triangle of dysplasia.¹⁷ The same regions often reveal fatty infiltration on black blood imaging. Less frequently observed findings include trabecular hypertrophy and wall thinning of the RV. In

patients with advanced ARVD, MR imaging may reveal left ventricular fat infiltration. This often occurs in the infero-posterior left ventricle and involves the epicardium of the left ventricle (Figure 1). Frank left ventricular dysfunction is unusual in early stages, but can occur in late stages of the disease and is an important reason for heart transplantation in ARVD.²³

Delayed enhanced MR imaging (DE-MRI) is very useful in demonstrating RV fibrosis, which in turn can guide the site for biopsy in ARVD.²⁴ Another important use of DE-MRI is in the differential diagnosis of ARVD, which will be discussed later in this article. The extent of delayed enhancement correlates with inducibility of ventricular arrhythmias during electrophysiologic testing.¹⁹ The anatomic origin of the ventricular arrhythmia can be predicted by the location of the fibrosis on DE-MRI, which can guide catheter ablation for VT. Shown in Figure 3A is a 12-lead ECG of a 24-year-old man who presented with wide complex tachycardia with a LBBB superior axis morphology. The morphology of the VT is consistent with its origin from the basal inferior right ventricle. DE-MR images (Figure 2) reveal fibrosis in the infero-basal right ventricle, the possible anatomic site of origin of the clinical VT.

Sarcoidosis. Sarcoidosis is a multisystem disease that is characterized by noncaseating granulomatous inflammation that involves predominantly the lymph nodes, lungs, musculoskeletal system, and the central nervous system. Cardiac involvement is seen in 5% to 25% of the cases and is an important cause of mortality.²⁵ Cardiac involvement often manifests itself as conduction disturbances and ventricular tachyarrhythmias. Diagnosis of cardiac sarcoidosis is often easy in the presence of

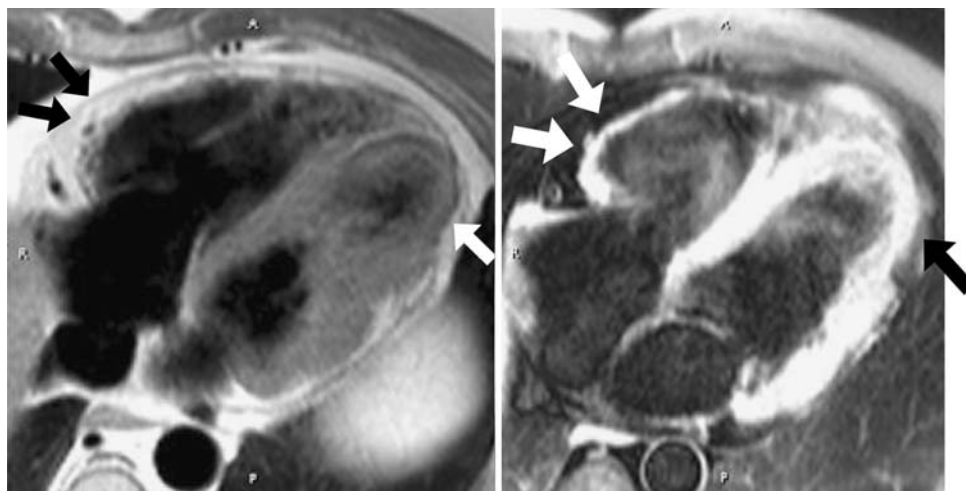


Figure 1. The left panel shows an axial black blood image of a patient with arrhythmogenic right ventricular dysplasia showing fat infiltration of the right ventricle (black arrows) and fat infiltration of the left ventricle (white arrow). Right panel is the same image with suppression of fat revealing the underlying epicardium (arrows), which appears thin.



Figure 2. A, Twelve-lead ECG showing left bundle superior axis ventricular tachycardia in a 24-year-old man with right ventricular dysplasia. B, Delayed enhanced MR image from the same patient showing hyper enhancement of the basal right ventricle, a region to which his VT was mapped during electrophysiologic testing.

multisystem disease; however, isolated cardiac sarcoidosis is not uncommon and is difficult to diagnose. Sarcoid granulomas or areas of myocardial scarring are typically present in the left ventricle and the interventricular septum in this condition. In some cases, the right ventricle can be predominantly affected raising the suspicion for ARVD.²⁶ A VT associated with RV abnormalities can, therefore, result in diagnostic confusion, especially if there is no systemic evidence of sarcoidosis. Appropriate diagnosis is very important as heart failure and arrhythmias in sarcoidosis respond to steroid therapy.^{25,27,28}

MR imaging, due to its high spatial resolution is the modality of choice to evaluate early stages of sarcoidosis. Myocardial inflammation caused by the granulomas

appears as high signal intensities in T1-weighted images. Studies have shown the utility of MR images to assess the benefit of steroid therapy as evidenced by resolution of the hyperintensities during follow up. Typically, the disease tends to involve the antero-septal region of the LV and the interventricular septum. Multiple discrete hyperintensities that are in a noncoronary distribution are highly suggestive of sarcoidosis.

Sarcoid involvement of the RV is characterized by RV dilatation and dysfunction, focal regions of akinesis, and aneurysmal dilation of the RV free wall. These findings are indistinguishable from either myocarditis or RV dysplasia. LV involvement is often present; however, it can be minimal or absent on imaging. Fat

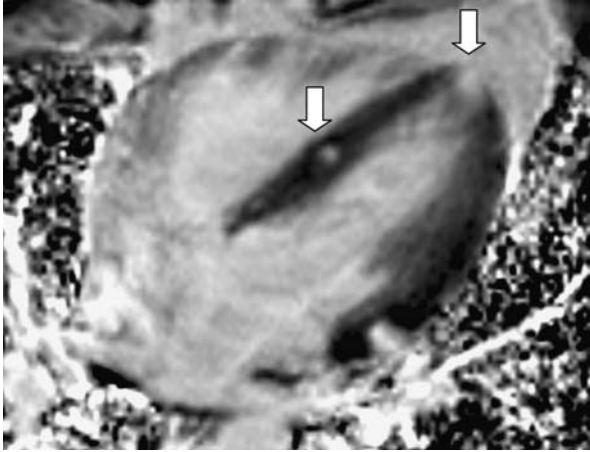


Figure 3. Delayed enhanced MR image from a 35-year-old man with sarcoidosis who presented with SCD. Focal enhancement of the interventricular septum, the apical left ventricle, and the lateral left ventricle are seen.

infiltration is usually not seen in sarcoidosis, and pulmonary hypertension is often an accompanying feature due to coexisting pulmonary disease.

Figure 3 shows a DE-MR image in a patient with sarcoidosis. Note the focal delayed enhancement of the interventricular septum and the apex and the lateral LV free wall. Pattern of myocardial delayed enhancement is extremely useful in differentiating sarcoidosis from ARVD and right-sided myocarditis. Although functional alterations are observed in the RV in sarcoidosis, patches of delayed enhancement in the LV are often present, which differentiates this from ARVD and isolated right-sided myocarditis. Delayed enhancement in sarcoidosis tends to be discrete and involves the full thickness of the ventricle, whereas delayed enhancement in ARVD spares the endocardium until the late stages. Finally, the majority of patients with sarcoidosis even without systemic manifestations have some degree of mediastinal lymphadenopathy that favors the diagnosis of sarcoidosis.

MR Imaging in Left Ventricular Arrhythmias

Coronary artery disease. Imaging is less useful in the diagnosis of CAD, as the diagnosis is usually apparent by history and by demonstrating evidence for diminished or absent coronary perfusion by direct angiography. The presence of a transmural or subendocardial scar in a coronary distribution on cardiac MR imaging may provide clues to the etiology. However, this is rarely used for diagnostic purposes. MR imaging may have a role in predicting inducibility of VT during electrophysiologic testing. Bello et al²⁹ studied 48 patients with known CAD who were referred for EPS using cine and

gadolinium-enhanced MRI. Infarct surface area and mass, as measured by cardiac MRI, were better identifiers of patients who have a substrate for monomorphic than LVEF. Furthermore, areas of scar on MRI has been shown to correlate with areas of low voltage on electroanatomic mapping, and identify critical targets of VT circuits during ablation in patients with prior myocardial infarction.³⁰ Infarct tissue heterogeneity on delayed enhanced MR imaging has been shown to be the strongest predictor of spontaneous ventricular arrhythmia with subsequent ICD therapy (as surrogate of SCD) among other clinical and MRI variables, in patients with previous myocardial infarction.³¹

Nonischemic cardiomyopathy. Apart from demonstrating decreased global function, which is non-specific, MR imaging can provide clues to diagnosis in nonischemic cardiomyopathies based on patterns of delayed enhancements. In contrast to CAD where the enhancement may be transmural, enhancement pattern in nonischemic cardiomyopathies tends to be isolated to the midwall or epicardium.³² Furthermore, the presence of delayed enhancement has been associated with adverse outcomes and inducibility of ventricular arrhythmias.

Hypertrophic cardiomyopathy. Myocardial hypertrophy in hypertrophic cardiomyopathy is easily demonstrated by MR imaging. Delayed enhancement of the RV attachment sites are frequently observed in this condition, but this finding is nonspecific and is seen in other conditions such as pulmonary hypertension, old age, and in conditions that lead to pathologic hypertrophy due to pressure overload. Anecdotal evidence suggests that the presence of patchy fibrosis confers a higher sudden death risk in HOCM patients.³³

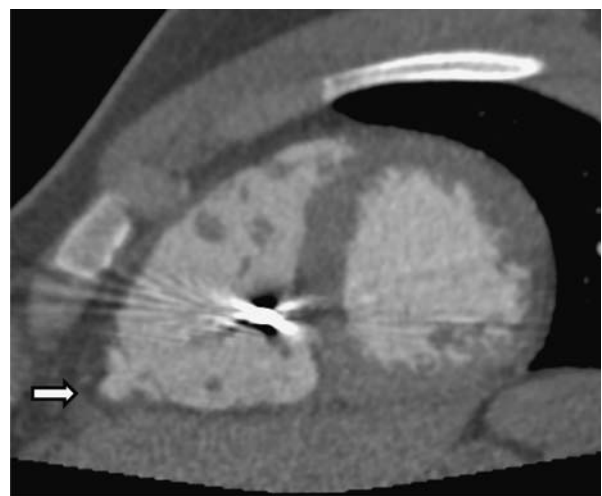


Figure 4. Right ventricular basal aneurysm demonstrated on reformatted CT images in a patient with sustained ventricular tachycardia.

ROLE OF CT FOR DIAGNOSIS OF ARRHYTHMIC SUBSTRATE

Computed tomography is extensively used in evaluating coronary anatomy in patients with intermediate risk for CAD; however, it is less frequently used for the purposes of cardiac morphology evaluation. CT imaging can similarly provide tissue characterization, identify intramyocardial fat deposits in ARVD, reveal mediastinal and pulmonary pathology in sarcoidosis, and depict

structural abnormalities associated with hypertrophic and dilated cardiomyopathies. Recent advances in multidetector row computed tomography have allowed imaging of myocardial scar in ischemic heart disease with a high spatial resolution.³⁴ Apart from the short image acquisition times, one distinct advantage of CT in arrhythmia evaluation is that a significant proportion of these patients have implanted devices which precludes MR imaging.

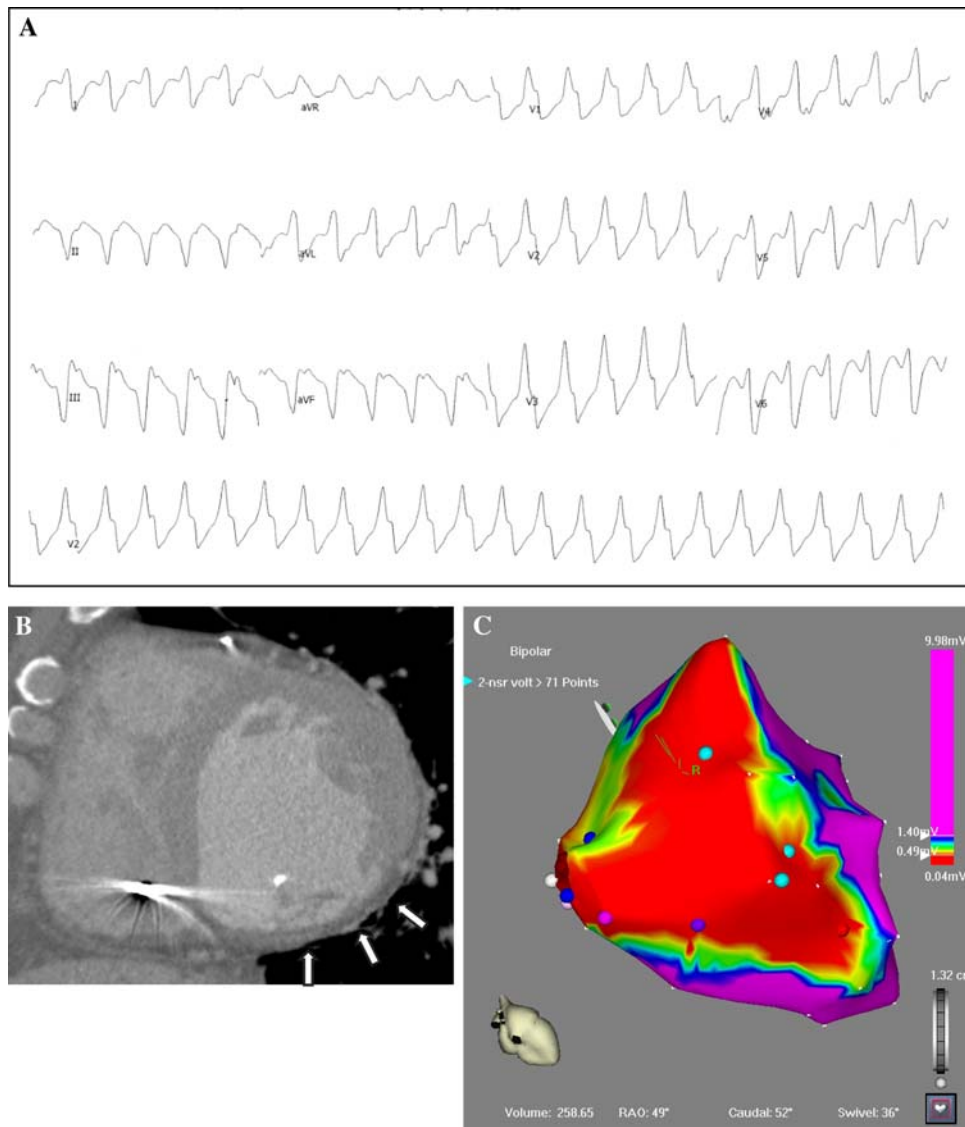


Figure 5. A, Twelve-lead ECG showing right bundle superior axis ventricular tachycardia in a 68-year-old man with a history of prior myocardial infarction. B, Short axis reformatted CT image shows a large inferior scar (arrows), an anatomic location consistent with VT morphology. C, Endocardial voltage map reveals a large area of low voltage in the inferior left ventricle, corresponding to the region of the scar. The ventricular tachycardia was mapped to the border of this scar with successful ablation.

MR AND CT IMAGING FOR LOCALIZATION OF ARRHYTHMIAS

Figure 4 shows an axial contrast CT image of a patient with ARVD who presented with sustained ventricular tachycardia. Note the discrete RV aneurysm in the lateral-basal right ventricle. During electrophysiologic study, the clinical VT was localized to the site of the RV aneurysm and was eliminated by radiofrequency ablation at this location. CT imaging in this patient with an implanted defibrillator provided clues to the location of the VT circuit and guided the electrophysiologic ablation as will be discussed later in this article. The

utility of CT and MR imaging in localizing the arrhythmias has been well established both in ischemic and nonischemic cardiomyopathies. Figure 5A shows 12-lead ECG of a patient with an extensive inferior wall myocardial infarction who presented with sustained RBBB inferior axis VT. CT image from the same patient shows extensive inferior scar. Large area of inferior low voltage was found during electroanatomic mapping and the VT was localized to the lateral boundary of the inferior infarct and was successfully ablated.

Bogun et al³² evaluated 29 patients with either VT or symptomatic PVCs with cardiac MR imaging and

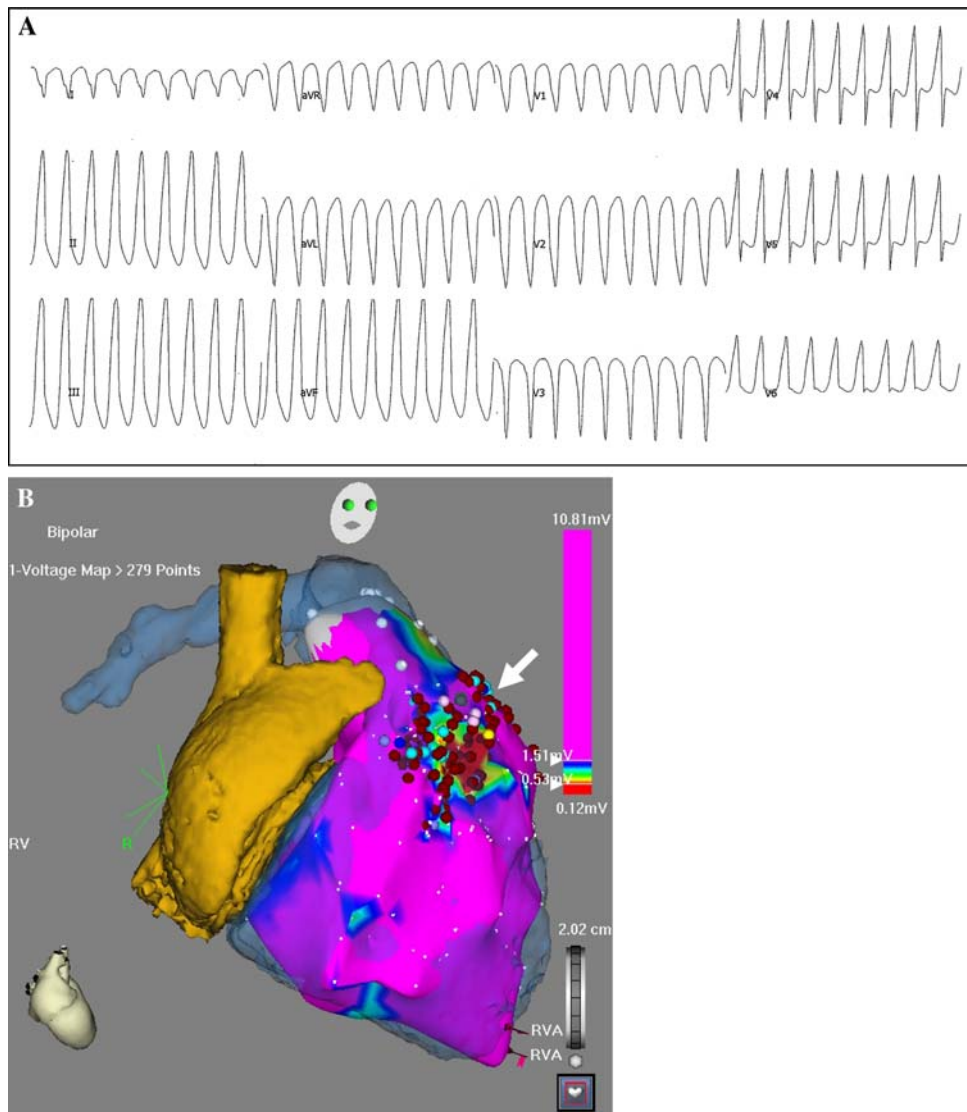


Figure 6. A, Twelve-lead ECG showing left bundle inferior axis ventricular tachycardia in a 28-year-old man with right ventricular dysplasia. B, Voltage map of the right ventricle merged on the CT image shows an area of low voltage in the right ventricular outflow tract (arrows). Red dots point to the region of radiofrequency ablation that led to success in eliminating the ventricular tachycardia.

classified scar morphology as either endocardial, epicardial, intramural, or transmural. Fifty percent of the study population had scar identified on MR imaging. In all of the patients with identifiable scar, the critical area for the VT during catheter ablation correlated with the region of the scar on MR imaging.

An important observation from this study was that, in patients with predominantly intramural delayed enhancement, catheter ablation was uniformly ineffective. Patients with predominant scar in the epicardium could not be ablated via an endocardial approach. Thus, the use of imaging prior to the procedure could provide valuable information that can help plan the approach for catheter ablation.

IMAGING TO GUIDE CATHETER ABLATION

Electroanatomic mapping involves image integration of electrical and pre-acquired anatomic maps during electrophysiologic procedures to enable catheter navigation. Anatomic maps are usually derived from CT or MRI images. This technique has revolutionized electrophysiologic procedures in terms of better understanding arrhythmia mechanisms, increased procedural success, and decreased complication rate.³⁵ Catheter navigation is traditionally guided by fluoroscopy, which provides only limited information about catheter location in relation to cardiac anatomy.

The ability to visualize the catheter tip in 3D in real time, in relation to a static electroanatomic map during electrophysiologic procedures, has significantly improved mapping of complex ventricular arrhythmias and outcomes of ablation in ventricular tachycardia.^{36,37} An example of such registration is shown in Figure 6. Figure 6A shows a 12-lead ECG of a patient with ARVD who presented with left bundle branch inferior axis VT. Shown in Figure 6B is the electroanatomic voltage map superimposed on the CT image revealing the location of the low voltage in relation to the 3D geometry of the right ventricle.

For high spatial resolution, CT is usually acquired using contrast enhancement and retrospective gating in axial orientation with 0.5 to 1 mm slice thickness. For MR imaging, MR angiography using nongated MR images (1 to 2 mm thickness) are obtained using a breath hold technique. Image registration is a crucial part of the integration process and refers to superimposing the 3D CT or MR image surface reconstructions onto the realtime electroanatomic maps derived from catheter mapping. Computerized registration algorithms are used to accomplish the image registration process. These algorithms are highly accurate and have been validated for clinical use.

FUTURE DIRECTIONS

Imaging has always been and will be crucial in the diagnosis and management of ventricular arrhythmias, and the role of imaging continues to grow. Contrast-enhanced techniques involving both MR imaging and CT imaging have found their application in tissue characterization and identification of anatomic substrates for nonidiopathic ventricular tachycardia. Mechanical activation maps by high temporal resolution anatomic images can further localize the site of arrhythmia in focal or triggered ventricular tachycardias. Investigations are under way to integrate realtime imaging techniques such as MRI and ultrasound with electroanatomic mapping, which provides realtime information on catheter-tissue contact and lesion formation.

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