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EDITORIAL COMMENT

Have We Reached the Apex?

Catheter Ablation in Hypertrophic Cardiomyopathy and Apical Aneurysm*

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Hypertrophic cardiomyopathy (HCM) is the most common genetically transmitted cardiomyopathy. It follows an autosomal dominant inheritance pattern, although it demonstrates variable expressivity, as well as age-related (and at times incomplete) penetrance (1). Even though the incidence of HCM has historically been estimated as in 1 in 500 individuals (2), more recent studies suggest that this number may be an underestimate (3). The spectrum of presentation in individuals with HCM is variable—patients may be asymptomatic, or they may have symptoms related to heart failure and arrhythmias. Indeed, patients with HCM have an increased incidence of supraventricular and ventricular arrhythmias, as well as sudden cardiac death (SCD).

In HCM, the most common hypertrophic site is at the confluence of the anterior septum and anterior left ventricular (LV) wall; this is associated with systolic anterior motion of the mitral valve that causes LV outflow tract obstruction. Two less common, yet important, phenotypes include apical HCM and HCM with apical aneurysm (AA). Although occasionally confused as 1, they are distinct entities. Patients with apical HCM have hypertrophy (≥15 mm) of the apical left ventricle. There is no LV outflow tract obstruction; however, patients may have midventricular obstruction (4). Although apical HCM is uncommon in Western countries, the incidence of this disorder is much greater in Far Eastern territories (5). Patients with apical HCM in the absence of an AA are generally at low risk for malignant arrhythmias, although atrial fibrillation is common (4). AA may be associated with apical HCM, but it may also occur independently. Aneurysmal development is believed to result from wall stress induced by the dynamic midventricular obstruction. However, given that some patients with AA do not have obstruction, other factors likely contribute, possibly including genetic predisposition, large-vessel coronary artery disease, microvascular disease, and midmyocardial coronary compression with associated hypertrophy (6,7). The supply-demand mismatch from the hypertrophy may lead to ischemia, which is worsened in the setting of increased intraventricular pressure during ventricular contraction. The AA itself is a discrete, thin-walled dyskinetic or akinetic segment in the distal portion of the left ventricle and may be composed of dense scar with channels of viable myocardium. Electrophysiologically speaking, the presence of an AA is important for 2 reasons: 1) it may form the nidus for endocardial thrombus; and 2) it may be the site of origin for rapid, monomorphic ventricular tachycardia (VT).

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To date, the characteristics and outcomes of patients with HCM, AA, and VT have been poorly defined and confined to isolated case reports or small case series (8,9). Given that patients with HCM and AA are at high risk for ventricular arrhythmias independent of conventional risk factors, they often

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receive an implantable cardioverter-defibrillator (ICD) for primary prevention. As such, although the incidence of SCD has decreased, patients may still present with appropriate ICD shocks. In this issue of *JACC: Clinical Electrophysiology,* Igarashi et al. (10) present a retrospective analysis of 15 patients with HCM and AA with monomorphic VT. In this patient cohort, Igarashi et al. (10) delineate the characteristics and outcomes of catheter ablation. Their main findings can be summarized as follows:

1. Of 15 patients, the average age was 65 ± 10 years. This is an interesting finding because although patients with HCM >60 years of age are typically thought to be at lower risk of SCD, those patients with AA may represent a unique cohort. Moreover, patients had a high prevalence of atrial fibrillation (40%).
2. Although only 6 of 15 patients had cardiac magnetic resonance (CMR) imaging, delayed enhancement was observed in the AA in all patients.
3. Baseline electrocardiographic abnormalities were commonly observed: T-wave inversion was seen in 12 patients, and ST-segment elevation was seen in 11 patients. The electrocardiographic pattern of the clinical VT was similar in patients: the majority had a north-west axis, and all had a QS pattern in the lateral precordial leads suggesting that the VT exit site was close to the LV apex. Of note, two-thirds had a right bundle branch block pattern in lead V_1, with the remainder having a left bundle branch block pattern, presumably dictated by whether the VT exit was from the free wall or the septal aspect of the aneurysm, respectively.
4. A total of 14 patients underwent 3-dimensional electroanatomic mapping, and CARTOSOUND (Biosense Webster, Irvine, California) was used in 6 patients. This illustrates that cases such as this require detailed intraprocedural imaging given the anatomic complexity. Even if CARTOSOUND is not used, intracardiac echocardiography is recommended to assist with catheter manipulation (especially through the aneurysmal neck), ensure adequate catheter contact, and promptly identify complications.
5. Endocardial substrate mapping and pace mapping were performed in all patients. Substrate mapping showed low-voltage and isolated late potentials within the AA in 80% of patients. Meanwhile, pace mapping was variably successful in identifying the exit site of the VT, presumably related to an intramural or epicardial exit. Entrainment mapping was possible in 6 patients, all with sustained, hemodynamically stable, monomorphic VT.
6. In 8 cases, the origin of the VT was presumed to be intramural or epicardial. Despite this, endocardial-only ablation was successful in 7 patients (open-irrigated catheter used). The remaining patient had only voltage and substrate abnormalities epicardially. Therefore epicardial ablation was performed. It is unclear what the endocardial unipolar signals demonstrated, and one would assume that because a true aneurysm is thin, endocardial ablation at the corresponding site would have been successful in terminating VT.
7. The rate of pericardial effusion was 7% (1 patient developed cardiac tamponade that was successfully managed with pericardiocentesis). One patient refused ICD insertion and had SCD 17 days after ablation.
8. A total of 80% of patients were free of VT during 12-month follow-up. However, the majority continued antiarrhythmic drug therapy (predominantly amiodarone) following their procedure. All patients received anticoagulation therapy following ablation, with no occurrence of thromboembolism.

As Igarashi et al. (10) highlight, the study was retrospective and had a small sample size (accruing only 15 patients across 5 centers over 10 years) and a relatively short follow-up. Further salient features should be highlighted:

Are this report and its findings applicable to everyday practice? The key issue is how many patients with HCM develop monomorphic VT and, more specifically, how many develop AA? In general, the incidence of sustained, monomorphic VT in patients with HCM is relatively uncommon. When seen, VT occurs in the context of myocardial substrate (secondary to myofibril disarray), post-myectomy scar, and AA. Indeed, ICD interrogations in patients with HCM typically show polymorphic VT degenerating into ventricular fibrillation (11). Further, the incidence of AA formation in patients with HCM is low and reported to be between 2% and 5% (7,12,13). Given that the overall incidence of HCM is <0.5%, the study findings are unlikely to be applicable to everyday practice. Nonetheless, they provide valuable insights, and Igarashi et al. (10) are to be congratulated for compiling a cohort across 5 institutions, although the small number of patients and multiple institutions has the potential to introduce significant heterogeneity into the study.

Can ablation techniques be generalized in patients with an AA? Most VTs in patients with AA arise within