Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is an inherited arrhythmic cardiomyopathy characterized by progressive fibro-fatty replacement of the right ventricular myocardium [1,2]. One of the major advances in the management of this life-threatening disease is the use of ICD (Implantable Cardioverter Defibrillator) to prevent sudden death as well as treat sustained ventricular arrhythmias [3]. The defibrillating lead is invariably placed in the right ventricle and may be affected by the progressive disease process, as such, long term lead performance in ARVC is of significant interest.

In this current issue of Indian Pacing and Electrophysiology journal, Lane et al. compared the lead parameters in ICD and CRT-D (Cardiac Resynchronization Therapy- Defibrillator) and indices of ventricular function among patients with Arrhythmogenic and Dilated cardiomyopathies (DCM) [4]. The authors conducted a retrospective record review of 1676 patients undergoing ICD or CRT-D implant at St Bartholomew’s and the Heart Hospitals, London between 2011 and 2016 and identified 18 patients with a confirmed diagnosis of ARVC who had a de-novo device placement. These patients were followed with a comparator group of 18 patients with confirmed diagnosis of idiopathic DCM for a mean of 30 months. Compared to DCM, ARVC patients started with a lower R wave amplitude at baseline (Δ −5.6 mV, p < 0.001), however, no significant change was observed in the lead parameters irrespective of septal versus non-septal locations. Similarly, RV lead threshold was significantly higher in the ARVC patients when compared to DCM group (Δ +0.2V, p = 0.031) which remain unchanged over time. There was limited data for LV lead threshold for ARVC patients as compared to the DCM patients who depicted significant increase with time. The authors also show a progressive decline in LV function in ARVC over 5 years with no change in DCM group. Data on TAPSE was restricted to ARVC patients which showed no significant change with time.

Contrary to prior reports [5], the authors found that the RV leads in the septal position had lower sensing during follow up compared to RV apical leads, which in fact had improved sensing. Right ventricular septum is characteristically spared in ARVC and often thought to be the best location for lead deployment due to its stable sensing parameters. The discrepancy in findings may be due to differences in the population and/or related to the small sample size and retrospective nature of this study. Progressive RV structural involvement would likely impact sensing in the RV apex which is usually affected in severe phenotypes. Another important finding of the study was the significant reduction of LVEF with progression in time in ARVC group as compared to the DCM group. Left ventricular involvement is well recognized in ARVC which may parallel or exceed the severity of right ventricular disease based on the genotype [6].

The results of this study add and extend our current understanding of ARVC disease progression and its impact on implantable devices. However, this study has to be interpreted with caution in light of its limitations. One of the main limitations is the retrospective study design where lead position was not controlled. Lead location at the time of the implant would be determined by the operator based on patient characteristics. It is quite possible that septal locations were specifically sought after in patients with severe RV phenotypes thereby influencing the final results. The small sample size also allows for variability in results that might not otherwise be explicable based on the pattern of RV disease progression in ARVC. Detailed phenotypic/genotypic information on the patients was unavailable which may be relevant to the findings. Patients with LV dominant ARVC are more similar to DCM and may have septal disease progression compared to conventional RV phenotypes. Another important information is the severity of RV disease and how it is related to both baseline RV lead parameters and lead function during follow up. Also, on closer look at the baseline demographics, the average mean age of the study patients is 51 years, which is a couple of decades older than the average ARVC population receiving implantable defibrillators. A previous study by Bhonsle et al. suggested that patients with late presentation (>50 years), though being at a similar arrhythmic risk as the early onset group, have significantly fewer arrhythmia events or appropriate ICD interventions, suggesting somewhat different and a less severe clinical course [7].

In addition to the theoretical concerns of worsening tricuspid regurgitation, the change in RV lead parameters is an important consideration in ARVC patients requiring transvenous implantable devices. Subcutaneous defibrillators overcome these limitations, but low R wave amplitudes and inappropriate interventions due to T wave oversensing have reduced enthusiasm for subcutaneous implants. For the foreseeable future, transvenous leads remain the first choice in ARVC patients as a good proportion of the sustained ventricular tachycardias are pace terminable. The authors’ efforts in identifying the changes in sensing parameters of ICD with the disease progression in ARVC patients is commendable and helps us to move a step ahead in the long-term outcomes of RV lead placement in this rare group of patients. These data are compelling and future studies with larger sample size of carefully phenotyped and genotyped patients with longer follow up duration are needed to confirm these findings.

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