The role of implantable cardiac defibrillators in cardiac sarcoidosis: saviour or sinner?

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This editorial refers to ‘Efficacy and safety of implantable cardiac defibrillators for treatment of ventricular arrhythmias in patients with cardiac sarcoidosis’ by J. Kron et al., on page 347

Sarcoidosis has been described as an enigmatic disease and shows significant heterogeneity in pattern, severity, and clinical course, with substantial geographical variations. For the majority affected, it carries a benign course, typically confined to the lungs and requiring no treatment.1 The same cannot be said when sarcoidosis involves the heart. Cardiac involvement in sarcoidosis manifests clinically in ~5% of patients, although autopsy studies have estimated cardiac involvement at closer to 30%.2 Progressive heart failure and malignant cardiac arrhythmias account for up to 77% of deaths due to sarcoidosis.3 Although there are no randomized studies evaluating the role of implantable cardiac defibrillators (ICDs) in cardiac sarcoidosis (CS), ICD implantation for CS carries a Class IIa recommendation in published guidelines.4

In this issue of the Journal, Kron et al.5 report on the largest retrospective series of patients with CS and ICDs. A total of 235 patients from Canada, USA, and India were included in their analysis, with a mean follow-up period of more than 4 years. Within the limitations of a retrospective analysis, their data reinforces two important points: first, CS has a high incidence of potentially life-threatening arrhythmias; secondly, ICDs are not benign devices.

Two other single-centre series assessing the long-term follow-up of patients with CS and ICDs have been published recently. Schuller et al.6 published a series of 112 patients with CS and ICDs, and Betensky et al.7 a series of 45 patients. Although some patients are acknowledged to feature in more than one series, the messages from each study are fairly consistent. The majority of patients (62–74%) received their ICD for a primary prevention indication, with a mean left ventricular ejection fraction of ~45%. The annual appropriate ICD therapy rate ranged from 8.6 to 15%, notably higher than seen in large primary prevention ICD trials, such as SCD-HeFT (Sudden Cardiac Death In Heart Failure Trial), where the rate was 5.1% per year.8

While it is evident that potentially life-threatening arrhythmias occur frequently in this patient population, the benefits of ICD therapy do not come without a price. Inappropriate shock therapy was seen in almost a quarter of patients (24.3%). While atrial fibrillation was the commonest cause, mechanisms to avoid this, such as a tailored approach to device programming require further evaluation. Other ICD-related adverse events occurred in ~15% of patients, most notably lead dislodgement or fracture. These factors need to be balanced against the potential benefits offered by ICD therapy and it is incumbent on physicians to openly discuss these issues with patients prior to implantation.

More effective risk stratification in patients with CS would potentially improve the risk: benefit ratio for ICD therapy. Programmed stimulation in CS has been shown to predict future arrhythmic events in two small series,9,10 although in one series 10% of patients with a negative ventricular stimulation study experienced sustained ventricular arrhythmia or sudden cardiac death during follow-up.10 All three case series following CS patients with ICDs found reduced ejection fraction to be a predictor of appropriate ICD therapy,5–7 although the majority of patients receiving therapy had an ejection fraction greater than the 35% cut-off used in many studies for primary prevention ICDs, suggesting this cut-off is not applicable for CS. Two of the studies also showed that advanced conduction system disease, represented by either complete heart block or ventricular pacing, was also a significant predictor of appropriate ICD therapy.5,7 This is, perhaps, not surprising as advanced conduction system disease is likely to be a surrogate marker for more extensive granulomatous infiltration of the myocardium and specifically the septum. It does, however, raise the question as to whether patients with sarcoidosis and standard pacing indications should be encouraged to have a primary prevention ICD implanted at the outset, rather than a simple pacemaker.

A further challenge in the management of patients with non-cardiac sarcoid is the issue of how to screen for cardiac involvement. While only a minority of patients will have cardiac sarcoid, it carries an adverse outcome and accounts for a significant
mortality in this population. There is very little guidance on how patients should be screened for cardiac involvement other than directed investigations when symptoms occur and utilization of a multidisciplinary approach to management of the patient with sarcoid.\textsuperscript{11}

In the absence of randomized studies of ICDs in CS (and it is unlikely any will be undertaken), the study by Kron et al. provides valuable insight into the role ICDs play in CS patients. Undoubtedly ICDs save lives, but not without exacting a price, so it is important that physicians and industry continue to refine device therapy so as to minimize inappropriate shocks and adverse events.

Conflict of interest: P.R.R is currently conducting research for Medtronic and St Jude Medical. He has received consultancy fees from Medtronic and Boston Scientific.

References