HEART FAILURE AND CARDIOMYOPATHY

Determinants of implantable defibrillator discharges in high-risk patients with hypertrophic cardiomyopathy

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Objectives: To identify the determinants of appropriate and inappropriate implantable cardioverter-defibrillator (ICD) discharges in patients with hypertrophic cardiomyopathy (HCM).

Design: Retrospective cohort study.

Setting: ICD clinic at an academic hospital.

Patients: 61 patients with HCM who received ICDs for the primary or secondary prevention of sudden cardiac death (SCD).

Outcome measures: (a) Analysis of appropriate and inappropriate ICD discharges; (b) predictors of ICD discharges.

Results: Mean (SD) age at ICD insertion was 46 (18) years (range 10–79). Follow-up time was 40 (27) months (range 7–151). Eight patients experienced an appropriate discharge, occurring 24.5 (13.6) months after ICD insertion. Appropriate ICD intervention was more common in the secondary (36%) than the primary (8%) prevention group ($p=0.02$). Inappropriate ICD discharges occurred in 20 (33%) patients. Multivariate Cox regression analysis identified two significant predictors of inappropriate ICD discharges: (a) age <30 years at the time of ICD insertion (hazard ratio (HR)=3.0 (95% CI 1.1 to 8.0; $p=0.03$) and (b) history of atrial fibrillation (HR=3.1 (95% CI 1.2 to 8.1; $p=0.02$).

Conclusions: ICDs are effective in the prevention of SCD in HCM. However, there is a high incidence of inappropriate ICD discharges.

Sudden cardiac death (SCD) is the most catastrophic complication of hypertrophic cardiomyopathy (HCM).¹ In patients with HCM there is a general consensus that patients who survive a cardiac arrest or sustained ventricular tachycardia (VT) should receive an implantable cardioverter-defibrillator (ICD) for secondary prevention of SCD.¹ ICD treatment may also be effective for primary prevention of SCD in high-risk patients.² Identification of patients at sufficient risk to warrant lifelong device treatment is a major challenge.¹ Furthermore, ICD treatment may be associated with adverse sequelae.² Unlike patients with ischaemic cardiomyopathy, many patients with HCM are young and may be exposed for decades to potential complications of an ICD. We aimed at identifying variables associated with appropriate and inappropriate ICD discharges in patients with HCM.

METHODS

This was a single-centre retrospective study of 61 patients with HCM followed up at the Toronto General Hospital. ICDs were inserted between 1996 and 2003 (except for two patients whose devices were implanted elsewhere and whose management was transferred to the Toronto General Hospital at age 18).

An appropriate discharge was defined as a discharge for termination of VT or ventricular fibrillation (VF). An inappropriate discharge was defined as a discharge without preceding VT/VF. Discharges were designated as appropriate or inappropriate by an electrophysiologist (DAC), based on morphological and R–R interval analyses and all available recorded diagnostics on stored electrograms. The VF detection threshold was 320 milliseconds.

The separate end points of first appropriate and inappropriate discharges were analysed using the Kaplan–Meier method. Groups were compared with the log rank test. A multivariate

Cox model identified predictors of inappropriate discharge. Statistical analyses were performed with SAS 8.2.

RESULTS

We studied 61 patients (40 male, mean (SD) age 46 (18) years (range 10–79), 44 dual-chamber) with HCM (mean (SD) maximal left ventricular wall thickness 22 (6) mm). Seventeen patients had a history of atrial fibrillation (AF). ICDs were inserted for primary prevention in 50 patients and secondary prevention in 11 (resuscitated cardiac arrest or sustained VT). Primary prevention patients were referred because of the presence of ≥ 1 major risk factor for SCD: non-sustained VT (33 patients), family history of premature (age ≤ 50 years) HCM-related death (28 patients), syncope (15 patients) and wall thickness ≥ 30 mm (9 patients).

One patient died suddenly during the follow-up period of 40 (27) months (range 7–151). This patient was a 22-year-old woman with recurrent appropriate discharges whose ICD was programmed to detect VT >120 /min. Review of the ICD showed no device malfunction and no detections of VT. Postmortem examination did not disclose any non-cardiac cause of death. Her death was presumably due to electromechanical dissociation or to undetected VT (<120 /min). One other patient underwent cardiac transplantation for refractory heart failure.

Nineteen appropriate ICD discharges were documented in eight patients (13% or 4%/year), occurring at 24.5 (13.6) months (range 4–44): 15 episodes triggered by VT/VF and four by atrial tachycardia (AT) or AF that degenerated into VT/VF. These four patients were younger (25.0 (19.4) vs 47.1

Abbreviations: AF, atrial fibrillation; AT, atrial tachycardia; HCM, hypertrophic cardiomyopathy; HR, hazard ratio; ICD, implantable cardioverter-defibrillator; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia

Figure 1 Kaplan-Meier curve for first inappropriate discharge in the cohort with hypertrophic cardiomyopathy. Inappropriate implantable cardioverter-defibrillator interventions occurred in 20 (33%) patients.

(16.7) years; $p = 0.03$) than the remaining cohort at the time of implantation. Primary and secondary prevention groups differed in freedom from appropriate discharges ($p = 0.02$): 8% (4/50) and 36% (4/11), respectively.

Inappropriate device treatment was the most common complication. Twenty patients (33% or 10%/year) had ≥ 1 inappropriate discharge, occurring at 15.3 (16.5) months (range 0.25–56) (fig 1). Two patients experienced both appropriate and inappropriate discharges. The first inappropriate discharge was accounted for by AF (eight patients), AT (three patients), sinus tachycardia (four patients), T wave oversensing (two patients), and lead fracture (three patients). Eight patients (13%) had a lead fracture or dislodgement and these patients were significantly younger than patients without lead complications (29 (14) vs 48 (17) years; $p = 0.006$).

Multivariate analysis identified two variables as significant predictors of inappropriate discharges: (a) age < 30 years at the time of ICD insertion (hazard ratio (HR) = 3.0 (95% CI 1.1 to 8.0; $p = 0.03$)) and (b) history of AF before ICD insertion (HR = 3.1 (95% CI 1.2 to 8.1); $p = 0.02$). Drugs had no significant impact on the development of inappropriate discharges.

DISCUSSION

Our rate of appropriate discharge (4%/year) in high-risk patients with HCM is similar to the rate of 4–7%/year detected in other studies of comparable HCM cohorts.^{2,3} Despite the benefits of ICD treatment, there were important adverse effects, and inappropriate ICD discharges occurred in 20 (33%) of our patients (10%/year).

Other studies of ICD treatment in patients with HCM have reported inappropriate interventions in about 25% of patients (5–8%/year).^{2,3} These discharge rates are generally higher than that reported in contemporary cohorts of non-HCM patients but comparable to that observed in children or adolescents with ICDs, where inappropriate discharges have been reported in up to 50% of patients.⁴ One of the significant determinants of inappropriate discharge in our study was young age at

implantation. In addition, because most of our patients had minimal exercise intolerance, it is plausible that they would be more prone to exercise-induced tachyarrhythmias than the older and functionally limited patients with significant left ventricular dysfunction who are typically offered ICDs.

Pre-existent AF was the other significant predictor for inappropriate intervention in our study, which has previously been reported with non-HCM patients.⁵ Conversely, there were also four cases of AT/AF, which precipitated VT/VF and triggered an appropriate discharge. The latter invokes a potential mechanism for SCD in patients with HCM and AF, and patients with HCM who develop AF are known to have a poorer prognosis.¹ The optimal care of patients with HCM, AF and an ICD should therefore include particular attention to the prevention of recurrent AF.

This study is limited in that it is a single-centre, retrospective study of a relatively small number of patients. Our cohort comprised patients managed at a tertiary referral centre and our results may not be applicable to all patients with HCM who receive ICDs.

ICDs are effective in the management of life-threatening arrhythmias in high-risk patients with HCM. Younger age and prior AF were independently predictive of inappropriate discharge. These observations are important considerations in the decision to offer prophylactic ICD treatment. With increasing numbers of patients with HCM receiving ICDs, this study highlights the need to refine the selection of patients, to determine appropriate ICD settings for this group and to optimise the management of AF in patients with HCM.

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Competing interests: None.

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