SCIENTIFIC LETTER

Outcomes after implantable cardioverter-defibrillator treatment in children with hypertrophic cardiomyopathy

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Implantable cardioverter-defibrillators (ICDs) have been shown to successfully treat life-threatening arrhythmias in high-risk adults with hypertrophic cardiomyopathy (HCM).[1] Children represent <1% of individuals with ICDs, and paediatric studies include few children with HCM. This study reports the experience with ICDs in children with HCM in a single referral centre.

METHODS

Between 1993 and February 2006, 160 consecutively referred patients with HCM (age ⩽16 years) underwent clinical evaluation using 12-lead ECG, two-dimensional, M-mode and Doppler echocardiography, Holter monitoring and cardiopulmonary exercise testing using previously described methods.[2] Risk markers for sudden cardiac death (SCD) were: (1) family history of SCD; (2) unexplained syncope; (3) abnormal blood pressure response during upright exercise; (4) non-sustained ventricular tachycardia; and (5) severe left ventricular hypertrophy (⩾30 mm).[2] Patients with previous cardiac arrest or with ⩾2 risk factors were considered for ICD implantation.[2] All patients who underwent ICD implantation during this period were included in this study.

Informed consent for ICD implantation was obtained from a parent in all cases. Devices were implanted under general anaesthesia into subpectoral pockets, using transvenous lead systems. Sixteen patients received dual chamber devices and six received single-chamber devices. Stored ICD data were obtained every 6 months or within 24 h of treatment. Discharges were judged appropriate when triggered by ventricular tachycardia or ventricular fibrillation, or inappropriate if triggered by sinus tachycardia, supraventricular arrhythmia or device malfunction.

The end points used in the survival analysis were: (1) SCD; (2) appropriate ICD discharge; (3) cardiac transplantation; and (4) death due to congestive heart failure. Data are presented as median (range) or mean (95% confidence intervals (CI)). Survival estimates, cumulative rates of appropriate shocks and annual discharge rates were estimated by the Kaplan–Meier method.

RESULTS

In all, 22 patients underwent ICD implantation. There were no deaths or transplantations. Mean follow-up to study end points or last follow-up (for those with no events) was 1.7(range 1–2.3) years for the whole cohort. Median follow-up was 1.4 years (mean 1.7 years, 95% CI 1 to 2.5) for the primary prevention group, and 10.1 months (mean 1.4 years, 95% CI −0.8 to 3.6) for the secondary prevention group.

Four (18.2%) patients received 15 appropriate shocks (3 patients in the secondary prevention group and 1 in the primary prevention group). In each case, a single shock successfully terminated the arrhythmia. Median time to first appropriate shock was 3.3 (range 1.4–10) months. Three patients were receiving β-blockers at the time of ICD discharge, three were receiving verapamil and two were receiving amiodarone. A total of 11 (73.3%) appropriate shocks were for ventricular fibrillation, 10 of which were preceded by sinus tachycardia (including the 2 shocks in the primary prevention patient). The remaining shocks were for polymorphic (n = 2) and monomorphic (n = 2) ventricular tachycardia. Annual discharge rates were 13% for the whole cohort, 71.4% in the secondary prevention group and 4.1% in the primary prevention group. The 5-year shock-free survival was 80.3% (62.9–97.9%) for the whole group, 93.3% (80.7–106%) for the primary prevention group and 40% (−2.9–82.9%) for the secondary prevention group.

In all, 4 (18.2%) patients had seven inappropriate ICD discharges (2 patients in each group). Median time to first inappropriate discharge was 1.2 years (13 days to 10 years). In three patients, inappropriate shocks were caused by sinus tachycardia or supraventricular tachycardia; further inappropriate discharges were prevented by using negatively chronotropic drugs (n = 2) or device reprogramming to prevent T-wave sensing (n = 1). The fourth patient had a discharge after lead fracture 10 years after ICD implantation.

One patient developed a haematoma at the implantation site. One patient experienced anxiety and depression associated with ICD discharges. One patient developed acute bacterial endocarditis (*Staphylococcus lugdunensis*) 6 months after ICD implantation, requiring emergency surgery. There was no obvious source of primary infection. She made a complete recovery and received a new ICD 3 months later.

DISCUSSION

Few studies have reported on the efficacy of ICDs in children with HCM. The largest paediatric ICD series was a survey of patients ⩽20 years old who received an ICD between 1980 and 1991, including 44 with HCM.[3] In all, 76% underwent device implantation for secondary prevention. The shock-free survival was 40% at 3 years, and 60% received appropriate discharges. However, devices at the time did not routinely allow storage of electrograms, making reliable classification of ICD discharges problematic. Furthermore, as most ICDs were implanted surgically, the results may not be applicable in the current era.

In our study, appropriate discharge rates in the secondary prevention group were higher than in adults (71.4% vs 11%).[1] This suggests that children with sustained ventricular arrhythmias have a substantially greater risk of further events than adults. The event rate in the primary prevention group was similar to adults (although only one patient had an event).

Abbreviations: HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter-defibrillator; SCD, sudden cardiac death

Table 1 Clinical characteristics at the time of implantable cardioverter-defibrillator implantation

	Whole group	PP	SP	p Value*
Demographics				
Male	13 (59.1)	8 (47.1)	5 (100)	0.05
Female	9 (40.9)	9 (52.9)	0	
Age at time of ICD implantation (years)	14 (7–16)	14 (12–16)	13 (7–16)	0.21
Age at diagnosis (years)	8 (0.3–16)	7 (0.3–15)	13 (2–16)	0.14
Symptoms and risk factors†				
Asymptomatic	4 (18.2)	2 (11.8)	2 (40)	0.21
Palpitation	9 (40.9)	9 (52.9)	0	0.05
NYHA dyspnoea class				
I	11 (50)	6 (35.3)	5 (100)	
II	8 (36.4)	8 (47.1)	0	0.04
III/IV	3 (13.6)	3 (17.6)	0	
Chest pain	12 (54.5)	11 (64.7)	1 (20)	0.14
Presyncope	10 (45.5)	8 (47.1)	2 (40)	1
Syncope	7 (31.8)	6 (35.3)	1 (20)	1
ABPR	15 (75)	13 (81.3)	2 (50)	0.25
FHxSCD	12 (54.5)	12 (70.6)	0	0.01
NSVT	1 (4.5)	1 (5.9)	0	1.0
Severe LVH	11 (50)	10 (58.8)	1 (20)	0.31
VF/polymorphic VT	5 (22.7)	0	5 (100)	NA
Drugs‡	16 (72.7)	14 (82.4)	2 (40)	0.1
Amiodarone	3	2	1	
β-Blockers	9	8	1	
Calcium antagonist	5	5	0	
Disopyramide	3	3	0	
Echocardiographic data				
MLVWT (mm)	29.5 (12–47)	31 (16–47)	20 (12–34)	0.12
Distribution				
ASH	21 (95.5)	17 (100)	4 (80)	
Concentric	1 (4.5)	0	1 (20)	0.23
LVEDD (mm)	39.5 (26–51)	39 (30–51)	42 (26–49)	0.98
LVESD (mm)	23 (10–34)	23 (10–34)	24 (15–30)	0.51
FS (%)	42 (31–67)	43 (31–67)	41 (33–45)	0.29
LAD (mm)	39.5 (29–56)	40 (29–56)	36 (29–43)	0.56
LVOTO ⩾30 mm Hg	8 (36.4)	7 (41.2)	1 (20)	0.61

ABPR, abnormal blood pressure response; ASH, asymmetric septal hypertrophy; FHxSCD, family history of sudden cardiac death; FS, fractional shortening; ICD, implantable cardioverter-defibrillator; LAD, left atrium diameter; LVEDD, left ventricular end-diastolic dimension; LVESD, left ventricular end-systolic dimension; LVH, left ventricular hypertrophy; LVOTO, left ventricular outflow tract obstruction; MLVWT, maximum left ventricular wall thickness; NSVT, non-sustained ventricular tachycardia; NYHA, New York Heart Association; PP, primary prevention; SP, secondary prevention; VF, ventricular fibrillation; VT, ventricular tachycardia.
Values are n (%) or median (range).
*PP vs SP.
†In the PP group, all patients had ⩾2 risk factors; 5 (29.4%) patients had 3 risk factors and 2 (11.8%) had 4.
‡6 patients were receiving >1 drug (amiodarone with β-blockers, n = 2; β-blockers with disopyramide, n = 2; β-blockers with calcium antagonists, n = 1; and disopyramide with calcium antagonists, n = 1).

It has been suggested that young children with HCM at high risk of SCD can be safely managed with amiodarone alone.[4] In our study, two of five patients receiving amiodarone experienced multiple appropriate ICD treatments. Although the numbers are small, this suggests that amiodarone does not prevent SCD in high-risk children.

Previous studies have highlighted the high incidence of inappropriate ICD treatments in the young,[5] resulting from higher heart rates at rest and during exercise. The rate of inappropriate discharges in our study is lower than in most childhood studies. The use of negatively chronotropic drugs and dual-chamber devices may explain this.

Previous reports have shown an increased frequency of complications compared with adults. In our study, only one patient had a wound complication. The youngest patient had psychological sequelae related to ICD discharges. The psychological effect of ICDs in young patients has been documented, and counselling and support should be available to patients undergoing ICD implantation.

One patient developed life-threatening infective endocarditis. As previous studies have documented a risk of ICD infection in young patients, there is a need for close vigilance against infection in this population.

The major limitation of this observational study is the small sample size. Nevertheless, this is the first study to examine the efficacy of ICDs in consecutively referred children with HCM who have undergone systematic risk stratification.

ICDs prevent SCD in high-risk children with HCM. Complication rates are lower than previously reported, but psychological support and prevention of infection and inappropriate shocks remain important issues.

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REFERENCES

1 **Maron BJ**, Shen WK, Link MS, *et al.* Efficacy of implantable cardioverter-defibrillators for the prevention of sudden death in patients with hypertrophic cardiomyopathy. *N Engl J Med* 2000;**342**:365–73.
2 **Elliott PM**, Poloniecki J, Dickie S, *et al.* Sudden death in hypertrophic cardiomyopathy: identification of high risk patients. *J Am Coll Cardiol* 2000;**36**:2212–18.
3 **Silka MJ**, Kron J, Dunnigan A, *et al.* Sudden cardiac death and the use of implantable cardioverter-defibrillators in pediatric patients. The Pediatric Electrophysiology Society. *Circulation* 1993;**87**:800–7.
4 **Seggewiss H**, Rigopoulos A. Management of hypertrophic cardiomyopathy in children. *Paediatr Drugs* 2003;**5**:663–72.
5 **Korte T**, Koditz H, Niehaus M, *et al.* High incidence of appropriate and inappropriate ICD therapies in children and adolescents with implantable cardioverter defibrillator. *Pacing Clin Electrophysiol* 2004;**27**:924–32.

IMAGES IN CARDIOLOGY

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True isolated right ventricular infarction with tombstone anterior ST elevation

A 73-year-old man presented with a 2-h history of ischaemic chest pain and dyspnoea. There was no antecedent history of ischaemic heart disease; however, the patient had been previously treated for hypertension. On presentation, he was haemodynamically stable with no clinical evidence of cardiac failure. His initial electrocardiogram (panel A) showed considerable ST elevation, particularly in the anterior leads.

The patient proceeded to primary percutaneous coronary intervention. The left coronary artery was free of marked disease. The right coronary artery was occluded in the proximal third. Restoration of flow after stent deployment showed a small-calibre, non-dominant right coronary artery (panel B), with resolution of the ST segment elevation. The patient developed transient hypotension, which improved with intravenous fluids, consistent with right ventricular infarction. Echocardiography showed normal biventricular function, reflecting the ability of the right ventricle to tolerate ischaemia.

Non-dominant right coronary artery occlusion may produce isolated right ventricular infarction; however, when associated with anterior ST segment elevation, it is usually seen in the context of concomitant inferior left ventricular dysfunction.

Dramatic anterior ischaemic ST segment changes complicating non-dominant right coronary artery disease is unusual, and, when seen, is usually associated with right ventricular and inferior left ventricular dysfunction.

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