Original Article

Role of Transvenous Implantable Cardioverter-Defibrillators in Preventing Sudden Cardiac Death in Children, Adolescents, and Young Adults

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• *Objective*: **To evaluate the indications, underlying cardiac disorders, efficacy, and complications involved with transvenous implantable cardioverter-defibrillators (ICDs) in pediatric patients at the Mayo Clinic.**

• *Patients and Methods*: **The records of all patients aged 21 years or younger who underwent transvenous ICD placement at the Mayo Clinic, Rochester, Minn, were reviewed retrospectively.**

• *Results*: **Between March 1992 and September 2000, 16 patients (7 females; mean age, 15.4 years; range, 10-21 years) underwent transvenous ICD placement. The ICD was implanted for primary prevention of sudden cardiac death in 7 and for secondary prevention in 9. The underlying cardiac disorders included hypertrophic cardiomyopathy in 6 patients and congenital long QT syndrome in 6 patients. The mean ± SD follow-up was 36±29 months (range, 5-108 months). There was no mortality. Seven patients (44%) received appropriate ICD therapy, including 6 of 9 who had ICDs placed for secondary prevention. Median time free from appropriate ICD discharge was 3 years (range, 0.2-9 years). Three patients (19%) experienced inappropriate ICD discharge. Two patients needed device replacement because of technical problems (lead fracture and device malfunction). Two patients developed pocket infection that required removal and reimplantation of the ICD.**

• *Conclusion*: **In adolescents and young adults, transvenous ICDs may prevent sudden death but are not free of complications. Forty-four percent of this cohort received potentially life-saving ICD therapy, including two thirds who received an ICD for secondary prevention.**

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ARVD = arrhythmogenic right ventricular dysplasia; DFT = defibrillation threshold; HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter-defibrillator; LQTS = long QT syndrome; OHCA = out-of-hospital cardiac arrest; QTc = corrected QT interval; VF = ventricular fibrillation

In the last 2 decades, the implantable cardioverter-defibrillator (ICD) has emerged as an important life-saving cardiac device. In the past, most clinical trials involving ICDs focused primarily on the adult population. In children, population-based studies[1] evaluating sudden death have reported an incidence of 1.3 to 8.5 per 100,000 patient-years, and many of the sudden deaths were attributed to a cardiac cause. Moreover, the survival rate for out-of-hospital cardiac arrests (OHCAs) has been poor in children.[2] Silka et al[3] reported that pediatric patients resuscitated from sudden cardiac death are at high risk for life-threatening arrhythmias. Therefore, the use of ICDs in young patients appears valid. However, in the past, their use in children and young patients has been limited, not only because of technical hindrances but also because of challenges faced in identifying the young person for whom ICD therapy is most appropriate.

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Implantable cardioverter-defibrillator technology has made immense progress in recent years. Initial designs for epicardial lead placement required surgical thoracotomy and were associated with more complications, high expense, and long hospital stays. With the advent of newer lead systems, a nonthoracotomy approach that uses transvenous leads became feasible. In addition to advancements in the leads, these devices have been downsized to allow subpectoral implantation. Even though these technological advancements have increased the applicability of ICDs to the pediatric population, there is limited knowledge regarding not only the procedural risks and complications but also the indications and benefits of ICD placement in this population.

The published experience with the use of transvenous ICDs in young patients is limited.[4-6] We report our experience in 16 young patients who underwent transvenous ICD placement. The implant indications, underlying cardiac disorders, efficacy, and complications involved with transvenous ICDs in these patients were analyzed.

PATIENTS AND METHODS

Records of all patients aged 21 years or younger who had a transvenous ICD placed at Mayo Clinic, Rochester, Minn, were retrospectively reviewed. Current status was obtained

by reviewing ICD or pacemaker clinic records or by contacting the local cardiologist where routine ICD interrogation occurred. Informed consent was obtained from each patient and/or the legal guardian for reviewing their records and obtaining follow-up information from their primary physician. This study was reviewed and approved by the Institutional Review Board of the Mayo Foundation. The pulse generators used were CPI Ventak models (Cardiac Pacemaker Inc, St Paul, Minn) in 10 patients and Medtronic models (Medtronic Inc, Minneapolis, Minn) in 6 patients (Table 1). The lead systems used included Endotek (Cardiac Pacemaker) in 10 patients and Transvene (Medtronic) in 6. The leads were positioned in the right ventricle with or without additional leads in the superior vena cava, right atrium, coronary sinus, or a subcutaneous patch electrode. After placement of the ICD, the lowest defibrillation threshold (DFT) was determined using a stepwise reduction in the energy delivered until failure and then testing it to ensure a safety margin of at least 10 J.

The underlying cardiac disease and indication for implantation were reviewed. The indication for device implantation was broadly classified as primary or secondary prevention. *Primary prevention* was defined as prophylactic implantation of an ICD in asymptomatic patients because of risk factors such as a positive family history of sudden cardiac death. *Secondary prevention* was defined as ICD placement in symptomatic patients who had experienced an OHCA or a sustained ventricular arrhythmia.

All appropriate and inappropriate discharges were noted. An *appropriate discharge* was defined as therapy delivered to terminate ventricular arrhythmia by antitachycardia pacing, synchronized cardioversion, or defibrillation. An *inappropriate discharge* was one that occurred during sinus or atrial tachycardia with heart rates just above the preset detection rate or due to myopotential oversensing. The duration of hospital stay after implantation was noted and all the complications were reviewed.

The time free of ICD discharge was determined by Kaplan-Meier analysis. Comparison of appropriate therapy in patients in whom ICDs were implanted for primary vs secondary prevention was done with use of a Fisher exact test. A P value of less than .05 was considered statistically significant.

RESULTS

From March 1992 to September 2000, 16 patients (7 females) received transvenous ICDs at the Mayo Clinic. The mean ± SD age at implantation was 15.4±3 years (range, 10-21 years), and the mean ± SD weight was 68.6±15.7 kg (range, 27.6-105 kg). Table 1 summarizes the underlying cardiac disorders and indications for ICD placement. The underlying cardiac disorders were cardiomyopathy (7), primary electrical disease (7), and repaired congenital heart disease (2).

Of the 7 patients with cardiomyopathy, 6 had hypertrophic cardiomyopathy (HCM) and 1 had arrhythmogenic right ventricular dysplasia (ARVD). The indication for an ICD in 5 of the 6 HCM patients was a family history of HCM and sudden death in a first-degree relative. The patient with ARVD presented with OHCA. The diagnosis of ARVD was proved by a myocardial biopsy specimen.

Of the 7 patients with primary electrical disease, 6 had long QT syndrome (LQTS) and 1 had presumed primary ventricular tachyarrhythmia. The 6 LQTS patients received ICDs because of OHCAs (2), breakthrough syncope despite β-blocker therapy (2), and a family history of LQTS and sudden cardiac death (2). The patient with primary ventricular arrhythmia presented with OHCA. He had a structurally normal heart with a normal myocardial biopsy specimen, a corrected QT interval (QTc) of 409 milliseconds, and no evidence of substance abuse.

Of the 2 patients with repaired congenital heart disease, 1 had aortic valve replacement with a mechanical valve for congenital valvular aortic stenosis. He received an ICD when he presented with OHCA. He later underwent a Konno procedure and developed complete heart block. The other patient had a complete repair of pulmonary atresia and ventricular septal defect and received an ICD after presenting with sustained ventricular tachycardia.

The ICD characteristics, including lead configuration and device model, are summarized in Table 1. The initial pulse generator was positioned in the left subpectoral region in 15 patients and in the abdominal position in 1 (case 14). All devices were biphasic except one, which was monophasic (CPI 1705). The mean ± SD DFT at implantation was 11.7±5.8 J (range, 3-20 J). Additional subcutaneous patch electrode (case 1) or extracardiac leads in superior vena cava (cases 2, 3, 14, and 15) were required in 5 patients because of initial DFTs exceeding 20 J.

The mean ± SD follow-up was 36±29 months (range, 5-108 months). There was no mortality. Seven (44%) of the 16 patients received appropriate ICD therapy on at least one occasion. Of the 9 patients in whom ICDs were implanted for secondary prevention, 6 received an appropriate ICD therapy compared with only 1 of the 7 in whom ICDs were implanted for primary prevention. However, this trend failed to achieve statistical significance (P=.06) because of small sample size. Of the 7 patients who received an appropriate discharge, the underlying pathologic condition was LQTS (4), HCM (1), repaired aortic stenosis (1), and ARVD (1). Two of the 4 LQTS patients and the patient with ARVD had multiple appropriate discharges (Table 1). Four of the 5 patients who presented with OHCAs as their sentinel event received appropriate ICD therapy (cases 7, 8, 9, and 15).

Table 1. **Clinical Profile of Transvenous ICD Recipients and Characteristics of Their Devices***

Patient No./ age (y)/sex	Underlying disorder	Indication	Lead configuration	Device model	DFT (joules)	ICD discharge		Antiarrhythmic therapy	
						A	I	Before ICD	With ICD
1/15/F	HCM	FH	RV:SQ	CPI 1705	20	1 VT	0	None	Amiodarone
2/20/M	HCM/ myectomy	FH	RV:SVC	Medtronic 7219	10	0	0	None	Atenolol
3/17/F	HCM/ myectomy	FH	RV:SVC	CPI 1743	5	0	0	Verapamil	None
4/10/F	HCM	FH	RV	CPI 1790	20	0	0	None	Atenolol, disopyramide
5/17/M	HCM/ myectomy	FH	RV	Medtronic 7229	3	0	0	Propranolol	Propranolol
6/10/M	HCM/ myectomy	VT	RV:RA	Medtronic 7271	6	0	0	Propranolol	Atenolol, amiodarone
7/17/M	ARVD	OHCA	RV	Medtronic 7223	20	>10 VF	0	None	Atenolol, amiodarone
8/16/F	LQTS	OHCA	RV	CPI 1763, 1851	10	4 VF	1 ST	None	Atenolol
9/18/F	LQTS	OHCA	RV	CPI 1763, 1793	15	1 VF	0	Atenolol	Atenolol
10/14/M	LQTS	Syncope with seizures	RV	CPI 1782	9	5 VF	5 ST	Propranolol	Propranolol, mexilitine
11/14/F	LQTS	Syncope with seizures	RV:RA	CPI 1851	6	4 VF	3 AT	Propranolol	Atenolol
12/14/M	LQTS	FH	RV:RA	CPI 1851	14	0	0	Atenolol	None
13/14/F	LQTS	FH	RV:RA	CPI 1851	5	0	0	None	None
14/21/M	Primary ventricular arrhythmia	OHCA	RV:CS:SVC	Medtronic 7217, 7221, 7227	15	0	0	None	None
15/14/M	Aortic stenosis	OHCA	RV:SVC	Medtronic 7223, 7221, 6945	15	1 VF	0	None	Propranolol
16/16/M	PA/VSD	VT	RV	CPI 1790	15	0	0	None	Atenolol

*A = appropriate; ARVD = arrhythmogenic right ventricular dysplasia; AT = atrial tachycardia; CPI = Cardiac Pacemaker, Inc; CS = coronary sinus; DFT = defibrillation threshold; FH = family history and sudden death in a first-degree relative; HCM = hypertrophic cardiomyopathy; I = inappropriate; ICD = implantable cardioverter-defibrillator; LQTS = long QT syndrome; OHCA = out-of-hospital cardiac arrest; PA/VSD = pulmonary atresia/ventricular septal defect; RA = right atrium; RV = right ventricle; SQ = subcutaneous; ST = sinus tachycardia; SVC = superior vena cava; VT = ventricular tachycardia; VF = ventricular fibrillation.

Median time free from first appropriate discharge was 3 years (range, 0.2-9 years; Figure 1). The patient with HCM who received an ICD for primary prevention received appropriate therapy 4 years after ICD implantation. This patient was defibrillated from sustained polymorphic ventricular tachycardia. Interestingly, 6 of 7 patients who received appropriate ICD therapy for ventricular tachycardia or fibrillation were taking β-blockers at the time of ICD discharge, and 2 were on combination drug therapy (Table 1).

Overall, 3 (19%) of the 16 patients experienced at least 1 inappropriate ICD discharge within 10 days to 14 months after device implantation (Figure 1). All 3 had LQTS and had also received appropriate ICD therapy. On interrogation of the device, sinus tachycardia in the ventricular fibrillation (VF) zone was found in 2 and atrial tachycardia in 1. After reprogramming the VF zone to higher heart rates, inappropriate discharges have not recurred in these 3 patients.

Of the 5 patients presenting with OHCAs, 4 had appropriate and 1 had inappropriate ICD discharge. Twelve patients were taking an antiarrhythmic drug after ICD implantation, including 4 who were on combination drug therapy (Table 1).

The mean ± SD duration of hospital stay after ICD implantation was 2.5±3.3 days (range, 1-14 days). Six patients underwent 1 or more replacements of their original ICDs because of end of battery life (3), lead fracture (1), device malfunction (1), need for dual-chamber pacing device (1), and pocket infection (2). The 2 patients who developed pocket infection required device removal and reimplantation. An 18-year-old woman with LQTS (case 9) developed pocket infection with *Staphylococcus aureus* 2 weeks after lead revision. The ICD and leads were removed, and she was treated effectively with antibiotics. A new ICD was implanted in the opposite pectoral region 2 weeks later. This patient was noted to have acne vulgaris of the face and trunk at that time. In addition, a 21-year-old man with primary ventricular arrhythmia (case 14) developed pocket infection 4 years after device implantation in the left upper abdominal quadrant. *Staphylococcus aureus* was isolated from the wound culture, and appropriate antibiotics were given. The ICD was explanted from the abdominal position and a new one placed 3 weeks later in the left subpectoral region.

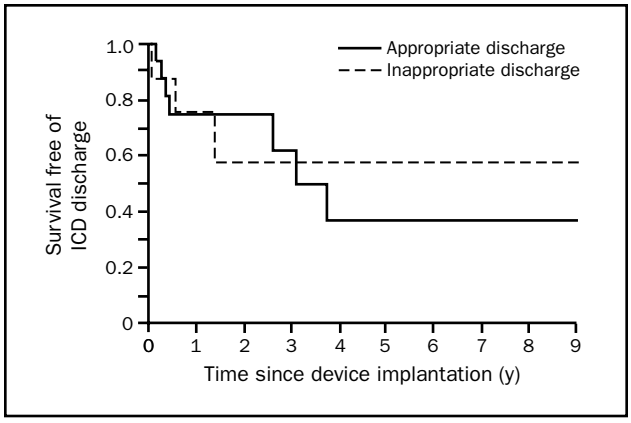

Figure 1. Time to first appropriate and inappropriate implantable cardioverter-defibrillator (ICD) discharge.

DISCUSSION

In children and adolescents, transvenous ICDs have proved to be effective in long-term prevention of sudden cardiac death in selected high-risk individuals in our study cohort. This predominantly adolescent cohort does not differ substantially with respect to body size from the adult population. Nevertheless, it is essential to define the role of ICDs in young patients because the causes of sudden cardiac death in children, adolescents, and young adults are different.

Sudden cardiac death resulting from tachyarrhythmias is less common overall in young patients than adults. However, specific pediatric populations are at high risk and their outcome is poor.[2] In adults, ICDs are effective in terminating life-threatening ventricular arrhythmias and saving lives.[7] In 1980, Mirowski et al[8] were the first to report the use of an ICD in humans. Since then, there has been a great revolution in the design and function of these devices. Advances have been made to reduce the size of battery and capacitor and achieve overall reduction in the size of the pulse generator.[9] With the advent of transvenous systems, experienced electrophysiologists are now implanting these devices in adults in the subpectoral or abdominal area without surgical assistance, with a high rate of success and few complications.[10]

Despite these advances, there is a paucity of published experience with the use of transvenous ICD systems in young patients and data regarding proper risk stratification for the underlying disorders, recommendations for ICD placement in young patients, and complications of the procedure itself.[4-6,11] In our study, the 2 most common underlying disorders for which an ICD was placed were HCM and LQTS.

The efficacy of ICD for primary and secondary prevention of sudden death in HCM was demonstrated in a recent multicenter study[12] involving 128 HCM patients (mean age, 40 years) with a high risk of sudden death. Of these 128 subjects, 85 had ICDs placed for primary prevention and 43 for secondary prevention. Overall, 29 (23%) of 128 patients received an appropriate shock, including 10 (12%) of 85 who underwent ICD placement for primary prevention and 19 (44%) of 43 who underwent ICD placement for secondary prevention. The interval between implantation and time of appropriate discharge was highly variable and prolonged in 6 patients (4-9 years). In our study, 5 of the 6 HCM patients underwent ICD placement because of a family history of sudden cardiac death (primary prevention). One of these patients (case 1) received an appropriate ICD therapy for ventricular tachycardia 5 years after the device was implanted.

The patient with ARVD (who presented with OHCA) has received more than 10 appropriate ICD therapies, despite concomitant treatment with amiodarone and atenolol. It has been shown that ICDs may confer a survival benefit of up to 50% in patients with ARVD.[13] In symptomatic LQTS patients, left untreated, mortality rates due to sudden cardiac death may be 20% in the first year and 50% at 10 years after onset of symptoms.[14] Moss et al[15] recently analyzed the effectiveness and limitations of β-blocker therapy in LQTS. This study, which involved 869 LQTS patients

from the International LQTS Registry, concluded that 32% of patients who had cardiac symptoms before starting β-blocker therapy will have another cardiac event within 5 years while undergoing β-blocker therapy. This provides evidence to support an ICD as first-line therapy when presentation of LQTS involves aborted cardiac arrest. In an international study of 287 children with LQTS, Garson et al[16] reported that 9% of these patients presented with cardiac arrest as their sentinel event. The authors suggested that in patients with extreme QT prolongation (QTc >600 milliseconds), especially if the patients continue to experience cardiac events while undergoing β-blocker therapy, consideration may be given to implantation of an ICD. Groh et al[17] reported that the use of ICDs in congenital LQTS is effective and safe, with low early and late complication rates. This study involved 35 LQTS patients, including 15 subjects aged 21 years or younger. It has been suggested that regardless of the degree of QT prolongation, ICDs should be considered for patients with LQTS presenting with OHCAs.[18] In our study, 4 of the 6 LQTS patients received potentially life-saving therapy within a median of 3 years of device implantation. All 4 underwent ICD placement as secondary prevention; OHCAs occurred in 2 and breakthrough cardiac events while undergoing β-blocker therapy occurred in 2. Notably, all 4 were receiving β-blocker therapy at the time of their shock and only 1 of these subjects had a QTc of more than 550 milliseconds. None of the LQTS patients in our study were treated with a combination of β-blockers and continuous pacing before ICD placement. Long-term follow-up studies[19] have failed to demonstrate a significant reduction in the risk for sudden cardiac death with such combination therapy.

Inappropriate shocks result from sinus or atrial tachycardia, with heart rates falling within preset detection zones or from false sensing of myopotentials. Myopotential oversensing may cause inappropriate shocks but was not seen in our study population. In this study, 3 LQTS patients received an inappropriate shock for sinus or atrial tachycardia. The interval between implantation and inappropriate shock ranged from 10 days to 14 months. The standard VF zone set for adult patients in our institution is 185 beats/min. Younger patients are able to achieve rapid heart rates with exercise. Consequently, the zone between sinus tachycardia and ventricular tachycardia or fibrillation may overlap. Keeping this in mind, reprogramming the VF zone to 220 beats/min may decrease the likelihood of future inappropriate shocks as seen in this series. Alternatively, dual-chamber ICDs may provide enhanced discrimination of sinus or supraventricular tachycardia from ventricular arrhythmia.[20] Two of these 3 patients had a single-chamber device and 1 had a dual-chamber device.

Although our adolescent cohort was adult-sized, the frequency of infections (12.5%) observed in this study was considerably higher than that seen for ICDs implanted in adults (0%-7%).[21] In our own institution, only 2 (0.3%) of 661 consecutive adult patients (average age, 63 years) undergoing nonthoracotomy ICD placement between September 1995 and August 2000 experienced a device-related infection requiring ICD removal (P. A. Friedman, MD, unpublished data). Further, a previous study[11] comparing complications in pediatric with adult patients also noted a higher incidence of device infection and lead revision in persons younger than 21 years. In that study, 2 (18%) of 11 adolescents had infection of the ICD system compared with 4 (1.3%) of 309 adults. In addition, 1 of 17 patients (mean age, 16.7 years) from a multicenter series developed a device-related infection.[4] Suboptimal wound care in younger persons has been speculated to underlie this higher infection rate.[11]

In our study, 2 patients experienced a device-related infection that necessitated explantation and placement of a new device at another site. First, an 18-year-old woman developed an *S aureus* pocket infection 15 days after lead revision. Interestingly, this patient had active acne vulgaris involving the face and thorax, a potential source of infection. A possible source of early pocket infections may be secondary to contamination from the patient's own skin flora.[21] The second patient developed infection late (4 years) after abdominal device implantation. A recently published large series[21] examining long-term infection rates in ICDs reported that infection was more frequent in primary abdominal implants compared with pectoral implants (3.2% vs 0.5%, respectively; P=.03). Abdominal ICDs may be more prone to infection because the procedure involves 2 stages in contrast to the single-staged pectoral implant and differences in local vascularity, cutaneous flora, and depth of adipose tissue.[21]

Besides infections, lead revisions may be more frequent in the adolescent population secondary to rapid growth and increased physical activity.[11] Previously, Link et al[11] found that lead revision necessitated by malfunctioning or migrated leads was done in 3 (27%) of 11 pediatric patients vs 33 (10.7%) of 309 adults. In our study, 1 adolescent (case 1) had a lead fracture 4 years after initial device placement that required lead revision. One might anticipate difficulties in vascular access in smaller pediatric patients, but this was not a problem in this series of essentially adult-sized adolescents. All patients had successful placement of leads via subclavian vein access. The duration of hospital stay for most patients was 1 to 2 days following the implantation except for patients who developed pocket infection.

Studies concerning psychological impact of ICD placement have shown that up to 38% of patients with ICD may

experience device-related anxiety and that young ICD recipients and those with high discharge rates experience the most psychosocial adjustment problems.[22] Restrictions on contact sports and driving can also have a marked impact on everyday life of adolescent and young adult patients. A recent study[23] addressing management of psychosocial issues in young ICD recipients has suggested screening and referral for psychological evaluation and to encourage an ICD "buddy system" to overcome social isolation and encourage effective coping in this group of patients.

Obviously, the retrospective review of a relatively small number of patients is a major limitation of this study. Because of this small cohort of adolescents and young adults, it is not possible to define any absolute guidelines for indications for ICD placement. Further studies are needed to better define the indications for ICD placement in young patients with cardiac disorders.

CONCLUSION

In this study, 44% of the adolescents and young adults who underwent transvenous ICD placement have already received potentially life-saving therapy from their device, including two thirds of the patients who had ICDs implanted for secondary prevention. The transvenous approach for ICD placement is safe and effective in young patients, although not free of complications. Transvenous ICDs are effective in aborting potential life-threatening arrhythmias and appear to play a critical role in secondary prevention of sudden cardiac death in young patients. The role of ICD therapy in primary prevention requires further investigation.

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