

Subcutaneous defibrillator implantation in pediatric patients

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ABSTRACT

Objective: Although sudden cardiac death is rare in children, an intracardiac defibrillator system is indicated in children with various types of cardiomyopathy, primary electrical diseases, and after surgical repair of congenital heart defects. The use of transvenous defibrillator lead systems is limited in pediatric patients because of a small body size and/or limited vascular access. Subcutaneous array leads combined with an abdominally placed generator can enable implantation.

Method: This is a retrospective study of 13 patients who underwent subcutaneous defibrillator implantation between September 2010 and March 2015. The subcutaneous system was preferred because patients were not amenable to transvenous lead placement.

Results: The median patient age was 4.1 years, and the median patient weight was 12.1 kg. Diagnoses of patients were long-QT syndrome in 6, aborted cardiac arrest with left ventricular non-compaction in 3, hypertrophic cardiomyopathy with sustained ventricular tachycardia in 3, and arrhythmogenic right ventricular cardiomyopathy in 1. Revision of the subcutaneous lead was required in 5 patients 2–26 months after the implantation. Appropriate shocks were observed in three patients. Inappropriate shock and lead fractures were observed in one patient during the follow-up period. The failure of therapy was observed in one patient. There were no perioperative complications and no early or late deaths.

Conclusion: Subcutaneous defibrillator systems are safe and effective in pediatric patients when the transvenous method is risky and contraindicated. Because the high growth rate in this population leads to lead failures, a close follow-up of this population is essential.

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Introduction

An intracardiac defibrillator (ICD) system is indicated for patients who are at a high risk of sudden cardiac death caused by ventricular arrhythmias. Sudden cardiac death is much rarer in children than in adults, occurring at an estimated incidence of 1–8 deaths per 100,000 patient-years (1). Children with various types of cardiomyopathy, primary electrical diseases, and after surgical repair of congenital heart defects are at a risk for sudden death caused by arrhythmia (2). ICD implantation is indicated in various age groups with varying size in a pediatric population.

The use of standard transvenous lead systems suitable for older children is limited in infants because of the small size. Although devices used for defibrillator implantation are getting smaller for pediatric patients, infants with lower body weights require specialized implantation techniques and devices (3). Subcutaneous array leads combined with an abdominally placed device can minimize the surgical approach and enable defibrillator implantation, particularly in patients in whom performing trans-

venous lead implantation has a high risk and is not appropriate.

Although this system seems to have advantages over transvenous systems with respect to implantation, multiple surgical procedures may be required during the follow-up of infants to adjust the electrode positions and/or revision of lead fractures. Also, there is no clear methodology for the implantation of an ICD in infants and small children because of the small number of patients; there is limited experience in this patient group (4).

The aim of this study was to evaluate the efficacy and safety of subcutaneous defibrillator systems implanted in children and report the results of midterm follow-up of patients.

Methods

Placement of the ICD system was performed in a cardiovascular operating room with the patient under general anesthesia. Bipolar epicardial lead was inserted at the right ventricle apex. Under fluoroscopy, the subcutaneous array lead was advanced downward and laterally to the back by blunt dissection using

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a metal tunneler and an introducer sheath between the 5th left intercostal area. Optimum sensing and pacing values were obtained during implantation. Defibrillation thresholds were determined by demonstrating successful conversions from induced ventricular fibrillation to sinus rhythm. Defibrillation shocks were delivered between the subcutaneous array lead as cathode and the “active can” ICD device as anode. All defibrillation systems had acceptable defibrillation thresholds.

During the follow-up period, threshold testing was not applied. The study protocol was approved by the locally appointed Ethics Committee.

Results

Between September 2010 and March 2015, 13 patients with indications for defibrillator therapy as primary or secondary prevention of sudden cardiac death underwent placement of a subcutaneous defibrillator system. The subcutaneous system was preferred in patients who were not amenable to transvenous lead placement because of small size, poor venous access, or having a previously implanted transvenous defibrillator with a history of infective endocarditis treatment and loss of venous access. Diagnoses of patients were long-QT syndrome in 6, aborted cardiac arrest with left ventricular non-compaction in 3, hypertrophic cardiomyopathy with sustained ventricular tachycardia in 3, and arrhythmogenic right ventricular cardiomyopathy in 1.

One patient had inducible ventricular tachyarrhythmia detected during the electrophysiologic study, and 6 had unexplained syncope related to inherited arrhythmogenic diseases. Two patients had pacemaker requirements because of an atrioventricular block, which developed after surgery for obstructive hypertrophic cardiomyopathy in one and was related with primary inherited disease in another patient.

At the time of the defibrillator system placement, the median patient age was 4.1 years, with a range of 1 month to 14 years. The median patient weight was 12.1 kg, with a range of 4–35 kg. The median follow-up period was 32.3 months, ranging from 3–58 months. There were no perioperative complications and no early or late deaths.

Three patients had previously implanted transvenous systems. The subcutaneous system was implanted after a median 3.5 years of follow-up because of infective endocarditis treatment in one patient and loss of vascular access in two patients.

Repositioning of subcutaneous lead was required in five patients (38%). Lead revision to achieve an electric field was performed 2–26 months after the implantation. The early revision requirement appeared in the pocket infection. Additional subcutaneous lead implantation was applied in three patients (Fig. 1).

Inappropriate shock due to lead fracture was observed in one patient during the follow-up period. Lead malposition leading to the failure of therapy was documented in one patient (Fig. 2). Successful therapy was applied with 35 J energy in that patient.

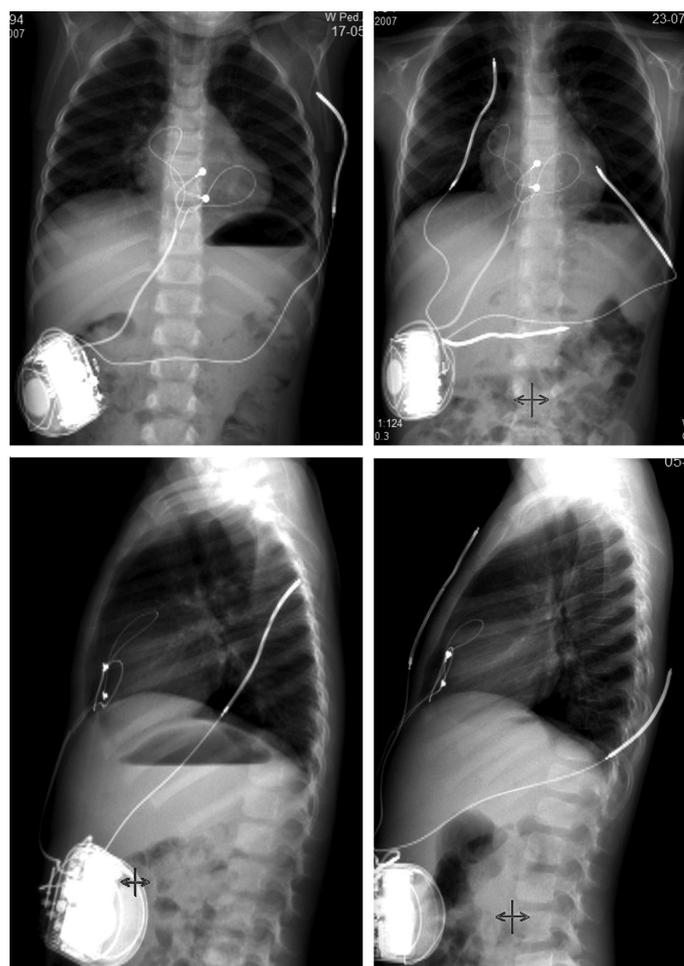


Figure 1. Revision of lead configuration by an additional lead 2 years after the implantation

All complications listed above were observed in one patient who had system implantation at the age of 16 months. Appropriate and successful shocks were observed in three patients (Table 1).

Discussion

The results of this study mainly demonstrated that the subcutaneous defibrillator system implantation in pediatric patients was effective in pediatric patients. However, a high lead revision percentage (38%) emphasizes the need of a close follow-up in this patient group with a high growth rate.

In children, life-threatening arrhythmia and ICD implantation are rarely observed. The indications for defibrillation therapy as primary prevention remains controversial, and the decision to implant an ICD in an asymptomatic child has often made it more difficult (5). Improvements in the risk stratification of diseases such as hypertrophic cardiomyopathy and recognition of genetic disorders such as long-QT syndrome and the number of pediatric patients selected for defibrillator implantation have increased over the years (4).

Improvements in implantation techniques and configurations have facilitated defibrillator implantation in young patients. Sub-

Table 1. Clinical characteristics of patients

Patient	Age	Weight	Diagnosis	Indication	Pacing mode	Lead revision	Appropriate shock
1	145	35	ARVC	Syncope	AAI/DDD		
2	24	11	LQTS	Syncope	VVI	yes	yes
3	35	15	LQTS	Syncope	VVI	yes	
4	1	4	LQTS, Complete AV block	Torsa Des pointes	VVIR	yes	yes
5	22	11	LQTS	Aborted cardiac death	VVI	yes	
6	108	16	HCMP	Sustained VT	AAI/DDD		
7	5	5	LVNC	Aborted cardiac death	VVI	yes	
8	110	17	LQTS	High risk	VVIR		
9	45	11	LQTS	High risk	VVI		yes
10	5	5	LVNC	Aborted cardiac death	VVI		
11	36	13	LVNC	Syncope	VVI		
12	60	12	HCMP	Sustained VT	VVI		
13	42	11	HCMP	Sustained VT	VVI		

ARVC - arrhythmogenic right ventricular cardiomyopathy; HCMP - hypertrophic cardiomyopathy; LQTS - long QT syndrome; LVNC - left ventricular non-compaction; VT - ventricular tachycardia

commonly reported, and fortunately, infection does not cause endocarditis, as can be seen in patients with endocardial leads.

Growth, particularly height, weight, and a change in body surface area were strongly associated with lead failure (12). Implantation of the subcutaneous system in older patients is associated with little or no lead revision requirement. Pettit et al. (14) reported no lead revisions when implantations were performed after 10 years of age. Because half of the patients were infants in the present study, the length and weight of these patients were small and all have high growth potentials. All lead revisions were required in patients in whom defibrillator implantation was performed below 3 years of age. No revision was performed above that age. The longer defibrillation coil of the subcutaneous lead compared with the shorter electrodes of the transvenous lead is more prone to accidents because of the high physical activity of patients. Although it has been thought that subcutaneous lead and abdominally-inserted active can is sufficient to establish a sufficient electrical field for defibrillation in the small chest of children, the mean defibrillation threshold at implant was higher (15.5 J) despite using various types of defibrillator implantation configurations compared with transvenous systems (11.5 J) (9, 15). A small displacement of the subcutaneous array may lead to an increase in the defibrillation threshold and failure of appropriate therapy.

All defibrillator system configurations have some advantages and disadvantages. In particular, when deciding non-transvenous system implantation, a decision should be individualized for each patient. An additional separate incision requirement for the placement of epicardial pacing-sensing electrodes and the generator seems to be a disadvantage of the subcutaneous system implantation. Intrapericardial implantation can be per-

formed through a single, upper abdominal incision without full or partial sternotomy. However, intrapericardial implantation may lead to pericarditis, life-threatening pericardial tamponade, and adhesions and may induce ventricular arrhythmias by irritating the myocardium. Implantation into a previously opened pericardium in operated congenital heart diseases is another limitation of intrapericardial implantation (16, 17).

Study limitations

The low ICD implantation rate in children leads to a small sample size. We furthermore acknowledge that this is a single-center experience, limited to subcutaneous defibrillator implanters. There have been limitations evaluating the advantages and disadvantages of applied therapies. Additionally, the course of the defibrillation threshold is unknown because of the loss of data collected for the efficacy of the system.

Conclusion

In our clinic, the subcutaneous systems were the method of choice for non-transvenous system implantation for avoiding pericardial complications. However, a close follow-up of these patients is essential because of commonly observed lead problems related with the growth of patients.

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