Circulation: Arrhythmia and Electrophysiology

ORIGINAL ARTICLE

The US Experience of the Wearable Cardioverter-Defibrillator in Pediatric Patients

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BACKGROUND: Certain pediatric patients are at risk for sudden cardiac death. The wearable cardioverter-defibrillator (WCD) can be used in clinical situations in which implantable cardioverter-defibrillator placement is not ideal. The objectives of the study are to examine the effectiveness, safety, and compliance of the WCD in the identification and treatment of life-threatening ventricular arrhythmias in pediatric patients.

METHODS: All United States pediatric patients <18 years who wore a WCD, from 2009 to 2016 were retrospectively reviewed.

RESULTS: In total, 455 patients were identified. The median age was 15 (3–17) years, median duration of WCD use was 33 (1–999) days and median patient wear time was 20.6 (0.3–23.8) hours per day. The population was divided into 2 groups: (1) patients with implantable cardioverter-defibrillator problem, n=63 and (2) patients with nonimplantable cardioverter-defibrillator problem, n=392. Wear time per day was >20 hours in both groups. Wear duration was shorter in the implantable cardioverter-defibrillator problem group, 26 days versus 35 days, $P<0.05$. There were 7 deaths (1.5%); all not wearing WCD at time of death. Eight patients (1.8%) received at least 1 WCD shock treatment. Of the 6 patients (1.3%) who had appropriate therapy, there were 7 episodes of either polymorphic ventricular tachycardia or ventricular fibrillation with a total of 13 treatments delivered. All episodes were successfully converted and the patients survived.

CONCLUSIONS: The WCD has overall adequate compliance with appropriate wear times and wear durations in pediatric patients. The WCD is safe and effective in treating ventricular arrhythmias that can lead to sudden cardiac death in pediatric patients.

VISUAL OVERVIEW: An online visual overview is available for this article.

Key Words: buffers ■ myocardial infarction ■ quality of life ■ syncope ■ ventricular fibrillation

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WHAT IS KNOWN?

- Certain pediatric patients are at risk for sudden cardiac death and there are patients that may not be immediate candidates for an implantable cardioverter-defibrillator.
- In adult studies, the wearable cardioverter-defibrillator can be used in patients that are not candidates for implantable cardioverter-defibrillators with survival >90%.

WHAT THE STUDY ADDS?

- The wearable cardioverter-defibrillator is commonly used in pediatric patients requiring implantable cardioverter-defibrillator repair or replacement along with higher risk populations with previous history of cardiac arrest of ventricular arrhythmias.
- The wearable cardioverter-defibrillator is safe and effective in treating ventricular arrhythmias that can lead to sudden cardiac death in pediatric patients.

Among populations that are known to be high risk for sudden cardiac death, the implantable cardioverter-defibrillator (ICD) has been shown to reduce mortality.[1,2] These high-risk patients may benefit from defibrillation protection acutely or chronically, but there are at risk patients in whom immediate implantation is either not feasible or not indicated. There are 2 groups of patients that may not be immediate candidates for an ICD (1) patients with a contraindication to immediate ICD placement or replacement (for example an infected device) and (2) patients who are at risk but may improve over time and not require long-term ICD therapy. The wearable cardioverter-defibrillator (WCD) is a therapeutic option for these patients and, in large adult studies, WCD safety and efficacy have been established.[3,4]

With an increasing number of studies demonstrating effectiveness of the WCD for treatment of ventricular arrhythmias, the American Heart Association recently released indications for WCD therapy in 2016. The recommendations for WCD therapy include (1) when a patient has a clear indication for an ICD but a transient contraindication occurs, (2) while awaiting cardiac transplantation, (3) during heightened risk of sudden cardiac death that may resolve over time with treatment of left ventricular dysfunction, and (4) during ICD waiting periods such as within 40 days of a myocardial infarction associated with increased risk of death in which ICDs have not been shown to reduce overall survival.[5] Just before this, in 2015 the Food and Drug Administration approved the WCD for use in children who weigh at least 41 pounds and have a chest size of at least 26 inches, which is about the typical size of an 8-year-old.[6] This has expanded the spectrum of patients for which the WCD may be considered.

ICDs are known to have relatively high rates of complications in the pediatric population, most commonly lead-related problems[7,8] and have a significant effect on the patient's quality of life.[9] For these reasons using a WCD instead of an ICD may be favorable in certain circumstances, particularly if the patient's condition may improve. There is limited data on the use of the WCD in pediatric patients.[10,11] The largest pediatric study previously published was by Collins et al,[11] in which there were no appropriate therapies in the patients ≤18 years old. The clinical experience with the WCD in adult populations is far more extensive. In large adult studies, the WCD has appropriate shock rates between 1% to 2% with acute survival >90%.[3,4,12]

We sought to evaluate the WCD use among pediatric-aged patients under age 18 from the manufacturer's US national database. Our objectives of this study were (1) to evaluate the WCD's ability to identify and treat life-threatening ventricular arrhythmias in pediatric patients and (2) to evaluate the effectiveness, safety, and compliance of the WCD in an up to date cohort.

METHODS

Because of the sensitive nature of the data collected for this study, requests to access the data set from qualified researchers trained in human subject confidentiality protocols may be sent to ZOLL LifeVest (Pittsburgh, PA).

Study Population

This was a retrospective study on consecutive pediatric patients <18 years of age who had a WCD prescribed by their physician. The study was approved by our institutional review committee. The patient data were provided by the ZOLL LifeVest (Pittsburgh, PA) maintained US registry. The registry includes demographic data, indication for prescription of the WCD (provided by the prescribing physician), related *International Classification of Diseases Ninth/Tenth Revision* code(s), length of use in days, daily wear time in hours (wear time per day), arrhythmias detected including the electrograms from the events, therapies provided, and patient-reported reasons for device return. The patients had to be <18 years of age before initial WCD placement and have worn the WCD for at least 1 day to be included in the study. We reviewed all patients that met these inclusion criteria from December 31, 2010, to September 14, 2016. The population was divided into 2 groups (1) patients who had the WCD placed because of an ICD problem (ICD problem) group and (2) patients with any other indication for the WCD (non-ICD problem) group. ICD problem was defined as patients with a previous ICD system with lead fracture, lead or system failure, and ICD infection. Non-ICD problem group included all other patients without an ICD problem.

Device Therapies

All therapies were reviewed and adjudicated by consensus of 3 electrophysiologists. A 2-lead electrogram of each therapy was available for review. Therapies were considered appropriate if delivered for ventricular tachycardia (VT) or ventricular fibrillation (VF). Therapy provided for any rhythm besides VT or VF was considered inappropriate. Successful therapies were defined as terminating the VT or VF. After screening out recordings that were noise, all nontreated recordings ≥60 seconds were also reviewed to find sustained arrhythmias that were not treated. Sustained arrhythmias were defined as 30 seconds or longer, but the minimum recording length for review was set at 60 seconds to accommodate the pre- and postdetection recording buffers of the WCD. All the therapy recordings for this study were ≥90 seconds, the minimal length of recording at which a therapy typically occurs. The reviewed recordings were defined as monomorphic or polymorphic VT, VF, sinus tachycardia, SVT or supraventricular tachycardia, or other by a qualified ECG technician.

Statistical Analysis

Because of the non-normal distributions of the continuous variables analyzed, medians with a range were used for reporting. Statistical analysis was performed using Stata 10.0 analysis software (Stata Corporation, College Station, TX). For the outcome variables comparing groups, univariate analyses were performed using the Wilcoxon rank-sum test for continuous variables and Fisher exact test for dichotomous variables. *P* values <0.05 were considered significant.

RESULTS

There were 455 patients who met inclusion criteria. Median age was 15 years (3–17), with 276 (61%) male. The median wear time per day was 20.6 hours (0.3–23.8) and days worn was 33 days (1–999; Table 1). Over half the population was 15 years of age or greater (Figure 1). There were a total of 185 centers that ordered a WCD and the median WCD ordered per center was 1 (1–30) with only 6 centers ordering >10 WCD (Figure 2). The median VT detection zone was 180 beats per minute (130–250) and VF zone 200 beats per minute (180–250). The median programming for shock therapy was 150 J (75–150) for all 5 shock therapies.

There were 63 patients classified as belonging in the ICD problem group, and of these, 36 (57%) had a mechanical problem with their ICD system and 24 (38%) had an infection (Table 2). Of the 392 patients in the non-ICD problem group, 167 (43%) had cardiomyopathy, 90 (23%) had congenital heart disease, 47 (12%) had channelopathies, and 36 (10%) had a cardiac arrest without another cardiac diagnosis. Before WCD placement 109 (28%) of all patients in the non-ICD problem group had a cardiac arrest and another 122 (32%) had a history of ventricular arrhythmia or concern for arrhythmogenic syncope; thus 231 (60%) had either cardiac arrest, ventricular arrhythmia, or concern for arrhythmogenic syncope (Table 3).

There were no differences in patient age, sex, or wear time per day between the ICD problem group and the non-ICD problem group. The wear duration in days was shorter in the ICD problem group compared with the non-ICD problem group, 26 days versus 35 days (*P*<0.05; Table 1).

Table 1. Patient Demographics

	Total Study	ICD Problem	Non-ICD Problem	*P* Value
Total patients	455	63 (14%)	392 (86%)	
Median age, y	15 (3–17)	15 (8–17)	15 (3–17)	NS
Male	276 (61%)	43 (68%)	233 (59%)	NS
Wear time per day, h	20.6 (0.3–23.8)	20.2 (0.7–23.4)	20.6 (0.3–23.8)	NS
Days worn, d	33 (1–999)	26 (1–415)	35 (1–999)	<0.05

ICD indicates implantable cardioverter-defibrillator; and NS, not significant.

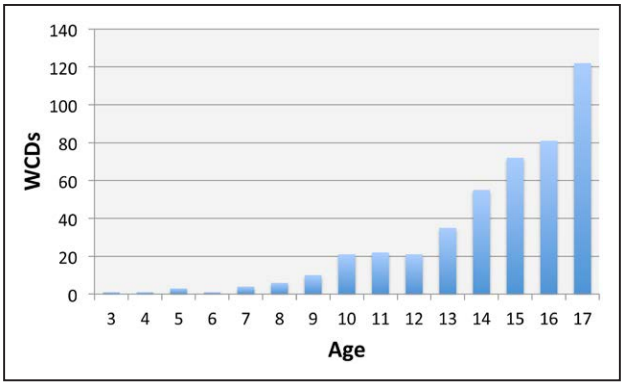

Figure 1. Wearable cardioverter-defibrillator (WCD) by age.
Total WCDs in the *y* axis with patient age (years) in the *x* axis. Over half the patient population was 15 y of age or older.

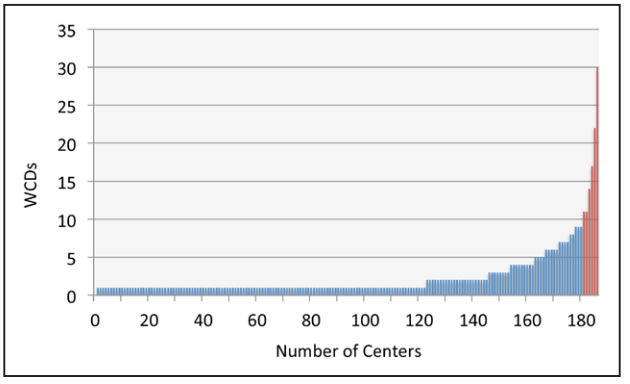

Figure 2. Wearable cardioverter-defibrillator (WCD) by center.
WCDs in number on the *y* axis. Number of centers on the *x* axis. Red columns are centers with >10 WCDs placed during the study period.

Table 2. Diagnosis for WCD (ICD Problem Group)

	N=63
Mechanical	36 (57%)
Lead fracture	12
Lead extraction	4
Lead dislodgement	2
Lead perforation	1
Lead revision	1
Infection	24 (38%)
Other	3 (5%)

ICD indicates implantable cardioverter-defibrillator; and WCD, wearable cardioverter-defibrillator.

WCD Therapies

There were a total of 8 patients (1.8% of the total study population) which received therapy from WCD. There were 6 patients with appropriate therapies (1.3% of the study population). There were 2 inappropriate therapies (0.4% of the study population; Table 4). The inappropriate therapies were secondary to oversensing of artifact during asystole (n=1) and noise/artifact during sinus rhythm (n=1). There were 3× as many patients 27 (6%) who personally aborted shocks during sustained arrhythmias. Most of these patients aborted therapies during sustained atrial/supraventricular arrhythmias 25 (93%) versus sustained ventricular arrhythmias 2 (7%). The median aborted episodes per patient were 2 (1–100). The median age for patients with appropriate therapies was 15.5 years (12–17), median wear duration 35 days (5–77) and wear time per day 21.3 hours (18.3–23). The median days worn to first shock therapy was 31 days (2–73). Of the 6 patients who received appropriate therapy, 2 had congenital heart disease (hypoplastic left heart syndrome variant and tetralogy of Fallot), 3 had cardiomyopathy, and 1 had a prior cardiac arrest. In a review of all appropriate therapies, there were 7 episodes of either polymorphic VT or VF with a total of 13 treatments delivered. Four of the 7 episodes terminated with the first treatment; 3 required 2 to 4 additional treatments. The median energy delivered by individual shocks was 150 J (150–154). In the 3 patients who required >1 shock to terminate the episode, all shocks were programmed for 150 J. All episodes were successfully converted and the patients survived (Table 5; Table I in the Data Supplement). In the ICD problem group, 0 of 63 (0%) received appropriate therapy compared with 6 of 392 (1.5%) in the non-ICD problem group; however, this result was not statistically significant (Table 4).

WCD Removal

The common reasons for WCD removal in the entire population were secondary to ICD repair or placement in 201 (44%), improvement of ejection fraction in 67 (15%), heart transplant/ventricular assist device in 20 (4%), and death in 7 (1.5%). A large percentage of the population 144 (32%) had the WCD removed for other reasons that

Table 3. Diagnosis for WCD (Non-ICD Problem Group)

	Non-ICD Problem	Cardiac Arrest	VT, VF, or Syncope
Total patients	392	109 (28%)	122 (32%)
Cardiomyopathy	167 (43%)	23	52
DCM	75		
HCM	34		
ARVC	8		
LVNC	8		
Other	42		
Congenital Heart Disease	90 (23%)	29	27
TOF	13		
TGA variant	12		
HLHS variant	11		
Anomalous coronary	9		
Aortic stenosis	8		
VSD	4		
Heterotaxy variant	4		
Shones	2		
Other	23		
Channelopathy	47 (12%)	11	23
LQTS	39		
Brugada	4		
CPVT	4		
Cardiac arrest (no other diagnosis)	36 (10%)	36	
Myocarditis	14 (4%)		
Syncope	12 (3%)		
Heart transplant	9 (2%)		
WPW	3 (<1%)		
Other	14 (4%)		

ARVC indicates arrhythmogenic right ventricular cardiomyopathy; CPVT, catecholamine polymorphic ventricular tachycardia; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; HLHS, hypoplastic left heart syndrome; ICD, implantable cardioverter-defibrillator; LQTS, long QT syndrome; LVNC, left ventricular noncompaction; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VF, ventricular fibrillation; VSD, ventricular septal defect; VT, ventricular tachycardia; and WPW, Wolff Parkinson White syndrome.

Table 4. Wearable Cardioverter-Defibrillator Therapies

	Patients (n=455)	ICD Problem (n=63)	Non-ICD Problem (n=392)	*P* Value
Therapy	8 (1.8%)	1 (1.6%)	7 (1.8%)	NS
Appropriate	6 (1.3%)	0	6 (1.5%)	NS
Inappropriate	2 (0.4%)	1 (1.6%)	1 (0.3%)	NS

ICD indicates implantable cardioverter-defibrillator; and NS, not significant.

Table 5. Appropriate Therapies

Total Patients (n=6)	
Demographics	
Age	15.5 (12–17)
Male	3 (50%)
Wear time per day, h	21.3 (18.3–23)
Days worn, d	39 (3–75)
Time to first therapy, d	31 (2–73)
Total episodes	7
Episode terminated first therapy	4 (57%)
Appropriate therapies	13
Arrhythmia (VT or VF)	13
Survived	6 (100%)

VF indicates ventricular fibrillation; and VT, ventricular tachycardia.

were not documented with a medical reason; within this group, 7 (5%) removed the device secondary to patient decision or noncompliance (Table 6). There were 7 patients who died (1.5%). Of those 7 patients who died, 4 were on milrinone therapy (2 were out of the hospital on milrinone) and 4 were in the hospital at the time of death. One patient had asystole out of the hospital and received an inappropriate shock was resuscitated and later died in the hospital. None of the patients that died in the study were wearing the WCD at time of death (Table 7; Table II in the Data Supplement). No patients who received appropriate therapy died during the study period.

DISCUSSION

This is the largest pediatric cohort to date demonstrating the US experience with the WCD. The study is also the first to describe appropriate therapy with a WCD in a pediatric population. There was considerable practice variability with >180 centers ordering at least one WCD but with 6 centers ordering 10 or more WCDs (Figure 1). Because pediatric-aged patients are unlikely to have a myocardial infarction, the demographics described in this study differ considerably from Chung's[4] adult US aggregate study with a higher percentage of patients with cardiomyopathy, congenital heart disease, and channelopathy. The majority of these patients (60%) were higher risk patients who had the WCD placed for either a previous cardiac arrest (28%) or a history of ventricular arrhythmia/concern for arrhythmogenic syncope (32%). The most common congenital heart diseases included tetralogy of Fallot, transposition of the great arteries, hypoplastic left heart syndrome variants, anomalous coronary arteries, and double outlet right ventricle variants. In this congenital heart disease group, 32% had a previous cardiac arrest and an additional 30% had a history of ventricular arrhythmia or concern for arrhythmogenic syncope.

Therapies occurred in 1.8% of the total population with appropriate therapies in 1.3% of the population. These numbers are consistent with adult studies in which appropriate therapies often occur between 1% to 2%. The WCD was 100% successful in terminating ventricular arrhythmia episodes (57% successful with first shock therapy) and all patients with appropriate therapies survived. The therapies in this cohort remain relatively small, therefore, creation of statistical models identifying risk factors for appropriate therapies was not performed. Of the 6 patients who received appro-

Table 6. WCD Removal

Reason for WCD Removal	Total Population (n=455)
ICD repaired or placed	201 (44%)
EF improved	67 (15%)
Heart transplant/VAD	20 (4%)
Condition deteriorates/hospitalized	9 (2%)
Death	7 (1.5%)
Cardiac surgery or catheter ablation	4 (1%)
AED	3 (1%)
Other	144 (32%)
Planned finish	32 (7%)
Equipment returned	29 (6%)
Patient decision	11 (2%)
Noncompliant	7 (2%)
Other	65 (14%)

AED indicates automatic external defibrillator; EF, ejection fraction; ICD, implantable cardioverter-defibrillator; VAD, ventricular assist device; and WCD, wearable cardioverter-defibrillator.

Table 7. Mortalities

	N=7 (1.5% Total Population)
Age	15 (10–17)
Male	4 (57%)
Wear time per day, h	20.3 (7–23.3)
Days worn, d	36 (5–218)
Diagnosis	
Congenital heart disease	3
Cardiomyopathy	3
Heart transplant	1
Therapy	
Asystole	1
Wearing WCD at time of death	0
Location at time of death	
Inhospital	4
Out of hospital (indications for removal include: removed for comfort, physician advised, no information)	3

WCD indicates wearable cardioverter-defibrillator.

priate therapies, 3 had cardiomyopathy (one with VT and another with VT and syncope), 2 patients with a previous cardiac arrest (one with hypoplastic left heart syndrome), 1 patient with tetralogy of Fallot awaiting heart transplant. Of these 6 patients, 4 (67%) had either a previous cardiac arrest or ventricular arrhythmia. Though 7 (1.5% of the study population) died, this group likely represented a sicker group of patients with 4 of 7 on milrinone and 4 of 7 patients in the hospital at time of death. None of the patients who died were wearing the WCD at time of death (exact cause of death in the 3 out of hospital arrest is not documented).

As previously demonstrated in adult studies, there are small percentages of patients (<1%) with a WCD that will have asystole, which can lead to death.[3,4,12] There was only 1 patient (0.2%) with asystole, a 17-year-old with a diagnosis of dilated cardiomyopathy with severe dysfunction who ultimately died after successful resuscitation. Currently, if there is concern that a patient is more likely to have asystolic cardiac arrest than VT/VF sudden cardiac arrest, then the WCD may not be the optimal therapy to prevent sudden cardiac death. Inappropriate therapies were uncommon, occurring only twice in the study (0.4%), one for oversensing of artifact during asystole and another for noise/artifact obscuring sinus rhythm. This percentage was consistent with adult WCD inappropriate therapies in the reported literature, ranging from 1% to 2%.[3,4,12] Because the WCD may oversense artifact and/or SVT or supraventricular tachycardia, the WCD has a warning mechanism with a noise alarm that allows patients to abort therapy if the patient wearing the WCD is asymptomatic and conscious. In the study, 6% of the patients had at least one personally aborted therapy during a sustained arrhythmia. The combination of low rates of inappropriate therapy and only a small percentage of patients that personally aborted therapy are likely important for patient compliance and their quality of life. Previous studies have shown that ICD therapy can lead to decreased quality of life in younger patients.[13]

Compliance with the WCD is obviously critical for detection and therapy for arrhythmias. There can be patient compliance issues, with common complaints in adult studies including weight of the monitor, skin rash/itching.[12] Median use of the device in adults has been reported to be >20 hours/day in adults[4] and just <20 hours/day in pediatric-aged patients. Wear time per day in our population was good and consistent with previous reports, with a median time of 20.6 hours/day.[11] Though the median days worn for the WCD was 33 days, there were patients in the cohort that wore the device for >1 year.

In the study cohort, just under half did not improve requiring either ICD repair or placement, heart transplant, or ventricular assist device placement. The most common reason for WCD removal was for ICD repaired or placement which occurred in 44% of the study population with another 4% either undergoing heart transplant or ventricular assist device placement. Fifteen percent of the population had improvement in their ejection fraction and the WCD was removed without an ICD.

Study Limitations

There were limitations for gaining clinical data from this registry, which was based on *International Classification of Diseases Ninth/Tenth Revision* codes along with a limited (when available) chart review for supplemental data. Also, there were a small number of therapies, which limited our ability to model risk factors for WCD therapies. A large percentage of the population had the WCD removed without clear medical reason available. Last, there were 3 deaths that occurred outside of the hospital, which could be considered failures as compliance may have factored into their deaths.

Conclusions

Pediatric patients overall had adequate compliance with WCD use, demonstrating appropriate wear times and wear durations similar to adults. The WCD is commonly used in patients requiring ICD repair and placement along with higher risk populations with either a previous history of cardiac arrest or ventricular arrhythmia/syncope. The WCD is safe and effective in treating ventricular arrhythmias that can lead to sudden cardiac death in pediatric patients. The rate of inappropriate therapies was low in this population. Pediatric patients were able successfully respond to prevent conscious shocks for ventricular and supraventricular arrhythmias. Additional comprehensive clinical data are required to understand which patients are at risk and may benefit from WCD therapy.

ARTICLE INFORMATION

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Disclosures

Dr Bianco is an employee of ZOLL. The other authors report no conflicts.

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SUPPLEMENTAL MATERIAL

Supplemental Table 1. Appropriate therapies patient descriptions

Patient	Age	Diagnosis	Time to first therapy (days)	Total episodes (n=7)	Initial arrhythmia	Total therapies (n=13)	Success
1	12	HLHS with cardiac arrest	9	1	VF/PVT	1	**yes**
2	17	DCM (FS 7%) with NSVT	73	1	VF	1	**yes**
3	17	DCM (EF 18%)	11	2	VF/PVT and PVT	1 and 3	**yes**
4	14	CM with syncope and inducible VT on EPS (EF 45%)	2	1	VF/PVT	2	**yes**
5	16	TOF with ventricular dysfunction (LV FS 27%) awaiting transplant	50	1	PVT	4	**yes**
6	15	Cardiac arrest	52	1	VF	1	**yes**

Supplemental Table 1. Appropriate therapies patient descriptions.

HLHS – Hypoplastic left heart syndrome; VF – ventricular fibrillation; PVT – polymorphic ventricular tachycardia; DCM – dilated cardiomyopathy; FS – shortening fraction; NSVT – nonsustained ventricular tachycardia; EF – ejection fraction; CM – cardiomyopathy; VT – ventricular tachycardia; EPS – electrophysiology study; TOF – tetralogy of Fallot; LV – left ventricle

Supplemental Table 2. Mortalities

Patient	Age	Sex	Wear time (hours)	Days worn (days)	Diagnosis	Therapy	Wearing WCD (time of death)	Location
1	17	F	21.4	26	DCM secondary to anthracycline (EF 30%) , trisomy 21 on milrinone	No	**No**	Out of hospital
2	11	M	7	25	VSD repair with DCM (EF 21%) on milrinone	No	**No**	Out of hospital
3	17	F	18.2	110	DCM (EF 10%) on milrinone	**Yes (asystole)**	**No**	In-hospital (resuscitated and alive when paramedics removed WCD
4	10	F	20.3	35	Heart transplant with coronary vasculopathy	No	**No**	In-hospital
5	15	M	23.3	218	Marfan with aortic root replacement, severe AR and heart failure (LV FS 7%)	No	**No**	Out of hospital
6	14	M	22.6	130	TGA/DORV s/p Fontan with NSVT on milrinone	No	**No**	In hospital
7	17	M	18.5	5	Cardiomyopathy (EF 32%) from Scleroderma with VF cardiac arrest	No	**No**	In-hospital

Supplemental Table 2. Mortalities

WCD – Wearable cardioverter defibrillator; DCM – dilated cardiomyopathy; EF – ejection fraction; VSD – ventricular septal defect; LV – left ventricle; FS – shortening fraction; TGA – transposition of great arteries; DORV – double outlet right ventricle; s/p – status post; NSVT – nonsustained ventricular tachycardia; VF – ventricular fibrillation