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Cardiac implantable electronic devices in adults with tetralogy of Fallot

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Abstract

Background—tachyarrhythmias and bradyarrhythmias affect 20%–50% of adult patients with tetralogy of Fallot (TOF) and some of these patients will require cardiac implantable electronic devices (CIED) such as pacemaker and/or internal cardioverter defibrillator.

Methods—the Mayo adult congenital heart Disease database was queried for patients with repaired TOF and history of CIED implantation, 1990–2017. The study objectives were: (1) determine the occurrence of device-related complications defined as lead failure, lead recall, device infection and lead thrombus; and (2) determine the occurrence and risk factors for defibrillator shock.

Results—there were 99 patients (age 46 ± 14 years and 66 (66%) men) with CIED, and the CIEDs were 41 (41%) pacemakers and 73 (73%) defibrillators. indication for defibrillator implantation was for primary prevention in 28 (38%) and secondary prevention in 45 (62%). Device-related complications occurred in 20 (20%) patients (lead failure 17, lead recall 4, device infection 12 and thrombus 3). Twenty-five per cent of all device infections occurred within 30 days from the time of device generator change. Annualised rates of appropriate and inappropriate shocks were 5.7% and 6.2%, respectively. The use of class III antiarrhythmic drug was protective against defibrillator shock.

Conclusion—the current study provides useful outcome data to aid patient counselling and clinical decision-making. Further studies are required to explore ways to decrease the risk of postprocedural infection after device generator change, and to determine which patients will benefit from empirical antiarrhythmic therapy as a strategy to decrease incidence of inappropriate defibrillator shock.

INTRODUCTION

Similar to the surgery for most congenital heart disease, surgical repair of tetralogy of Fallot (TOF) is a palliative procedure and most patients have residual or recurrent haemodynamic

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lesions or arrhythmogenic substrates.¹² These haemodynamic lesions or arrhythmogenic substrates determine clinical outcomes, and interventions are often required to address these residuals in order to improve clinical outcomes.^{3–9} Tachyarrhythmias and bradyarrhythmias affect 20%–50% of adult patients with TOF and some of these patients will require cardiac implantable electronic devices (CIED) such as pace-maker and internal cardioverter defibrillators.^{810–13}

Although the use of CIED may provide symptomatic and survival benefits in some patients, these devices carry some associated morbidities such as device infection, inappropriate shocks, thromboembolism, mechanical injury and need for multiple reinterventions with its associated procedural risks.^{812–14} There are several studies about outcomes of CIED in patients with congenital heart disease but data derived specifically from adults with TOF are rather limited.^{812–14} Therefore, the purpose of this study was to describe outcomes of CIED in adults with TOF.

METHODS

Patient selection

The Mayo Adult Congenital Heart Disease (MACHD) database was queried for patients (age 18 years) with repaired TOF and history of CIED implantation who had follow-up at Mayo Clinic Rochester Minnesota from 1 January 1990 to 31 December 2017. The patients with pulmonary atresia were excluded.

Data Collection

The following electronic health records were reviewed in detail: device implantation procedure note, device interrogation reports, clinical notes, ECG, Holter monitor and transthoracic echocardiograms. Other clinical variables were retrieved from the MACHD database.

Study Design

The study objectives were: (1) describe the clinical characteristics of patients with TOF with CIED; (2) determine the occurrence of device-related complication (DRC) defined as lead failure, lead recall, lead thrombus or device-related infection (generator pocket infection, bacteraemia with or without vegetation on device lead); and (3) determine the occurrence and risk factors for device therapy (appropriate shock and inappropriate shocks) in patient with implantable defibrillators.

Lead failure was defined as the presence of any of these features: failure to capture, failure to sense, abnormal pacing impedance and electrical noise artefact.¹⁴ For the analysis of the risk factors for defibrillator shock, we divided the patients into two groups: primary prevention group and secondary prevention group (defined as history of aborted sudden cardiac death and/or sustained ventricular tachycardia), and we tested the same risk factors as previously described by Khairy *et al.*⁸ Pacing modes were classified into: single-chamber atrial pacing (AAI), single-chamber ventricular pacing (VVI), dual-chamber pacing (DDD), biven-tricular pacing and biventricular pacing with defibrillator (CRT-D). In order to

evaluate for any temporal change in the incidence of DRC, we divided the study period into early era (1 January 1990 to 31 December 2006) and late era (1 January 2007 to 31 December 2017).

Statistical Analysis

Data were presented as mean $\pm$ SD, median (IQR) or number (%). Unpaired t-test, Wilcoxon rank-sum test, χ^2 and Fisher's exact test (as appropriate) were used to compare between-group differences. The risks factors for appropriate and inappropriate shocks were analysed using Cox proportional hazards model and expressed as HR and 95% CI. In the analysis of freedom from lead failure, we compared right atrial pacing leads versus right ventricular pacing leads versus defibrillator leads, using right atrial pacing leads as the reference. Coronary sinus leads were excluded from this analysis because of the small number of patients with these leads in the cohort. In the analysis of freedom from DRC, we counted only one event per patient, and the earliest DRC from the time of initial CIED implantation was selected for analysis. Kaplan-Meier's method and log rank test were used for these analyses. All statistical analyses were performed with JMP software (V.13.0; SAS Institute, Cary, NC). A $p < 0.050$ was considered statistically significant.

RESULTS

Baseline Characteristics

We identified 99 patients with TOF with CIED implantation out of 465 patients with TOF in the MACHD database (21%). There were significant differences in the baseline clinical and haemodynamic characteristics of the study group (TOF with CIED) and the rest of the TOF cohort (tables 1 and 2). The age at the time of initial TOF repair was 8 (4–15) years, the age at the time of initial CIED implantation was 46 ± 14 years and 66 (66%) were men. Of the 99 initial CIED implantations, 41 (41%) were pace-makers and 58 (59%) were defibrillators. The entire cohort was followed for 8.2 ± 7.5 years.

Pacemakers

Among the 41 patients with pacemaker implantation, the most common indication for pacing was high-grade heart block ($n=26$, 63%), and 35 (85%) patients received dual-chamber (atrial-ventricular sequential) pacing systems (table 3). The pacing modes at the time of initial implantation were AAI ($n=2$, 5%), VVI ($n=4$, 10%), DDD ($n=34$, 83%) and biventricular pacing ($n=1$, 3%), and 26 (63%) patients had rate responsive pacemakers. The left ventricular ejection fraction at the time of pacemaker implantation was $49\% \pm 12\%$.

A total of 17 (42%) patients subsequently underwent the following device upgrades: upgrade to atrial-ventricular sequential pacing ($n=2$, 5%); upgrade to dual-chamber defibrillator ($n=13$, 32%); and upgrade to CRT-D ($n=2$, 5%). Additionally, 13 (32%) patients had at least one pacemaker generator change, and the average longevity of a pacemaker generator was 8.4 ± 3.2 years.

Defibrillators

A total of 73 patients underwent defibrillator implantation, and 58 of these were initial CIED implantations while the other 15 patients received defibrillators during device upgrade of previously implanted pacemaker systems. The indications for defibrillator implantation were for primary prevention in 28 (38%) patients and secondary prevention in 45 (62%) (table 3). The left ventricular ejection fraction at the time of defibrillator implantation was $52\% \pm 11\%$. Electrophysiology studies were performed in 32 (44%) patients prior to defibrillator implantation. Of these 32 patients, 16 (50%) had inducible ventricular tachycardia, and ablations of ventricular tachycardia substrate(s) were attempted in nine patients of which six were successful (defined as absence of inducible ventricular tachycardia). The 16 patients who underwent defibrillator implantation without inducible ventricular tachycardia had other risk factors for sudden death such as left ventricular ejection fraction $<35\%$, prior syncope and significantly elevated left and/or right ventricular end-diastolic pressures.

The initial defibrillators implanted in these 73 patients were single-chamber systems ($n=18$, 25%), dual-chamber systems ($n=48$, 66%) and CRT-D ($n=7$, 10%). A total of 3 (4%) patients underwent subsequent device upgrades. One patient was upgraded from a single-chamber to dual-chamber system because of inappropriate shocks due to atrial tachycardia, and two patients were upgraded from dual-chamber system to CRT-D because of left ventricular dysfunction. Additionally, 26 (36%) patients had at least one generator change, and the average longevity of the device generator was 6.4 ± 2.7 years.

The defibrillator was reprogrammed as follows: (1) antitachy-cardia pacing zone for rate of 184 (171–193) beats/min; (2) slow ventricular tachycardia zone for rate of 196 (187–221) beats/min, and device was programmed to detect 19 (16–28) beats, redetect 14 (10–16) beats and to deliver one to two bursts of antitachycardia pacing and an initial shock with 35 (35–41) J; (3) ventricular fibrillation zone for rate of 214 (200–238) beats/min, and device was programmed to detect 19 (16–28) beats, and to deliver a burst of antitachycardia pacing and an initial shock with 35 (35–41) J. In four patients, an additional fast ventricular tachycardia zone of 235 (219–250) was also programmed in addition to the three zones described above.

The 73 patients with defibrillators were followed for 8.1 ± 7.3 years (594 patient-years), and there were 33 appropriate shocks (5.7% per year) in 30 (41%) patients and 37 inappropriate shocks (6.2% per year) in 34 (45%) patients. The mean detected ventricular tachyarrhythmia rate was 229 ± 63 , and 84% were monomorphic ventricular tachycardia while 16% were polymorphic ventricular tachycardia or ventricular fibrillation. There was no significant difference in the ventricular tachyarrhythmia rate between the primary and the secondary prevention groups ($p=0.314$). There were 19 cases of antitachycardia pacing of which 8 (42%) were successful in terminating the ventricular tachyarrhythmia. The 11 (58%) cases of unsuccessful antitachycardia pacing were all terminated by a single appropriate shock. In comparison to the primary prevention group, there was no significant difference in the rate of appropriate shock (5.5% vs 6.0% per year, $p=0.135$) and inappropriate shock (5.9% vs 6.4% per year, $p=0.097$). Of note, none of the 16 patients whose electrophysiology study was negative for inducible ventricular tachycardia had appropriate shock in contrast to 5 of the 16 patients (31%) with inducible ventricular tachycardia ($p=0.043$). Of the five patients

with history of inducible ventricular tachycardia prior to defibrillator implantation, two had successful ablation of ventricular tachycardia foci at the time of electrophysiology study. Table 4 shows the univariable risk factors for appropriate and inappropriate shocks. The use of class III anti-arrhythmic therapy was associated with lower risk of appropriate shock (HR 0.14, 95% CI 0.04 to 0.54, $p=0.004$) and inappropriate shock (HR 0.39, 95% CI 0.12 to 0.72, $p=0.008$).

Outcomes

A total of 242 leads (241 endocardial and 1 epicardial leads) were implanted and of these, 91 (38%) were right atrial leads, 54 (22%) were right ventricular pacing leads, 11 (5%) were coronary sinus leads and 85 (35%) were defibrillator leads. The epicardial lead was excluded from analysis. There were 17 (7%) lead failures and 4 (2%) lead recalls. The freedom from lead failure was 79% in 10 years (figure 1), and was similar between right atrial pacemaker leads and right ventricular pacemaker leads versus defibrillator leads 91% vs 89% vs 72% at 10 years, $p=0.395$.

There were 12 cases of device-related infection in 11 (11%) patients, and of these, nine cases were bacteraemia (*Staphylo-coccus* $n=4$, *Streptococcus* $n=3$, *Enterococcus* $n=2$) without vegetation, and two of these patients had concomitant generator pocket infection. There were three cases of bacteraemia (*Staphylococcus* $n=3$) with vegetation on the leads. Of these 12 cases, 7 (58%) were treated with intravenous antibiotics and device extraction (including the two patients with concomitant generator pocket infection), while 5 (42%) were treated with intravenous antibiotics alone. The five patients who received intravenous antibiotics alone were alive at the time of last follow-up. Of these device-related infections, 4 (33%) occurred within 30 days of generator change, and one patient subsequently died from sepsis and multisystem organ failure.

A total of 3 (3%) patients had lead thrombi (in the absence of bacteraemia or evidence of systemic infection) based on transthoracic echocardiogram, and all thrombi were attached to the right atrial lead. Two of these patients were on warfarin but had subtherapeutic anticoagulation (international normalised ratio <2.0) at the time of thrombus diagnosis while one patient was not on any anticoagulation. Intensive anticoagulation was initiated and therapeutic anticoagulation was achieved in all cases. Lead thrombus completely resolved in one of the patients while the other two patients showed reduction in the size of lead thrombus. The composite endpoint of DRC occurred in 20 (20%) patients, and the freedom from DRC was 74% in 10 years (figure 1). There was no significant difference in the annual incidence of DRC between the early and late eras (1.31% vs 1.18%, $p=0.153$).

There were 13 cases of lead extraction, and the indications for lead extraction were infection ($n=7$), lead failure ($n=4$), device upgrade ($n=1$) and extraction during cardiac surgery ($n=1$). Of these 13 lead extractions, 10 were performed in the electrophysiology laboratory while three were performed in the cardiac operating room. There were no procedural complications. Another 14 patients underwent additional lead implantation without lead extraction.

During the total duration of follow-up, 13 (13%) patients died and 2 (2%) patients underwent heart transplantation. The causes of death were end-stage heart failure (n=3), sudden cardiac death (n=2), sepsis/multisystem organ failure (n=1), stroke (n=1) and unknown (n=6). The two cases of sudden cardiac death occurred outside our hospital. The first patient was a 36-year-old man with prior pacemaker implantation for sinus node dysfunction and no history of documented ventricular tachycardia who died in his sleep at home. Autopsy was performed but the autopsy report was unavailable in his medical records. The second patient was a 52-year-old man with documented ventricular tachycardia and subsequent defibrillator implantation who had a witnessed syncope and appropriate defibrillator shock. The patient was transported to an outside hospital where he had refractory ventricular arrhythmia with multiple shocks and subsequently expired. The exact detail of events around the time of his death was not available in his medical record.

DISCUSSION

In this retrospective study we described outcomes in a cohort of 99 adult patients with TOF with CIED, of which 41 (41%) and 58 (59%) received pacemakers and defibrillators at the time of initial CIED implantation. Another 15 patients had device upgrade of previously implanted pacemakers to defibrillators resulting in a total of 73 patients with defibrillators. The indications for defibrillator implantation in the current study were consistent with the expert opinions expressed in the guidelines for management of adults with congenital heart disease.¹⁵ According to the guidelines, defibrillator implantation can be considered, in addition to therapy for residual haemodynamic lesions, in adults with repaired TOF in the setting of left ventricular systolic or diastolic dysfunction, non-sustained ventricular tachycardia, QRS duration >180 ms and extensive fibrosis on cardiac MRI.

The composite endpoint of DRC occurred in 20 (20%) patients, and the freedom from DRC was 74% in 10 years. Of these DRCs, 17 were lead failure, 11 were device-related infection and 3 were lead thrombi. The results of the current study are consistent with a previous multicentre study of 121 adult patients with TOF with implantable cardioverter defibrillator, which reported DRC in 25 (20%) patients of which 14 were lead failure/dislodgement, 1 case of endocarditis and 1 case of lead thrombus.⁸ This multicentre study did not provide further data about occurrence of bacteraemia and other device-related infection other than endocarditis. Another important observation from the current study was that 25% of all device-related infection occurs within 30 days after device generator change. Bacteraemia and endocarditis are major concerns in adults with congenital heart because of presence of risk factors such as prosthetic valves.¹⁶ The presence of transvenous leads further increases the risk of infection.¹⁷ In large prospective study from the Danish Pacemaker Registry that studied outcomes of 46 299 patients with CIED, the risk of device-related infection was about 1% per year and was significantly higher after device intervention such as a generator change similar to our study.¹⁷ It is difficult to pinpoint the specific factors related to generator change that are responsible for postprocedural infection because of the retrospective study design. An in-depth analysis of the procedural factors that predispose to infection is an important issue that deserves further study and maybe an opportunity for quality improvement.

The annualised rates of appropriate and inappropriate shocks were 5.7% and 6.2%, respectively, without any difference between the primary and secondary prevention groups. The results of the current study were comparable to previously reported annualised rates of appropriate and inappropriate shocks of 7.7% and 5.8%, respectively.⁸ The risk factors for appropriate shock were non-sustained ventricular tachycardia and elevated left ventricular end-diastolic pressure in the Khairy *et al*'s cohort. The current study did not identify any risk factors for appropriate shock, but the use of class III antiarrhythmic drug was protective against appropriate shock. The novel finding in the current study was the potential role of using inducible ventricular tachycardia for risk stratification considering the fact that none of the patients with negative electrophysiology study had any appropriate shock during follow-up. The small sample size and the fact that most patients did not undergo electrophysiology study prior to CIED implantation prohibited more robust statistical analysis to rigorously determine if the absence of inducible ventricular tachycardia was truly a marker of a low-risk subset of patients.

Limitations

This is a retrospective single-centre study, and by design had several limitations such as referral and recall bias. Although the study identified the use of class III antiarrhythmic drugs as a potential protective strategy against defibrillator shock, the study did not perform in-depth analysis to determine the specific types, dosage and duration of use of class III antiarrhythmic therapy that provided the most protective benefit. Additionally, less than half of the patients underwent electrophysiology study, which limited our ability to reliably determine if the absence of inducible ventricular tachycardia was a marker of a low-risk patient subset.

CONCLUSIONS

The study described the clinical characteristics of adult patients with TOF with CIED. DRC occurred in 20% of the cohort, lead failure occurred in 7% of the cohort while device-related infection occurred in 11%, and one-quarter of all infections occur within 30 days from the time of device intervention. The annualised rates of appropriate and inappropriate shocks were 5.7% and 6.2%, respectively, and were similar regardless of whether the device was implanted for primary versus secondary prevention. The use of class III antiarrhythmic drug was protective against appropriate shocks. The current study provides useful data to help in patient counselling and clinical decision-making. Further studies are required to explore ways to decrease the risk of postprocedural infection after device generator change, and to determine which patients will benefit from empirical antiarrhythmic therapy as a strategy to decrease incidence of appropriate shock.

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Key messages

What is already known on this subject?

- Tachyarrhythmias and bradyarrhythmias affect 20%–50% of adult patients with tetralogy of Fallot and some of these patients will require cardiac implantable electronic devices (CIED) such as pacemaker and/or internal cardioverter defibrillator. However, there are limited clinical outcome data about CIED in this population.

What might this study add?

- Device-related complications (lead failure, lead recall, device infection and thrombus) occurred in 20%. Device infection was most common within 30 days after device generator change. Annualised rates of appropriate and inappropriate shocks were 5.7% and 6.2%, respectively.

How might this impact on clinical practice?

- The study provides useful outcome data to aid patient counselling and clinical decision-making, and also supports further studies to explore ways to decrease risk of postprocedural infection after device generator change, and the incidence of inappropriate defibrillator shock.

Figure 1.
(Top) Freedom from lead failure; (bottom) freedom from device-related complication.

Table 1

Baseline characteristics

	All (n=465)	CIED (n=99)	no CIED (n=366)	P values
Age at beginning of study (years)	37±14	42±12	35±14	<0.001
Male (%)	223 (48)	66 (66)	155 (43)	<0.001
Body mass index (kg/m ²)	27±6	27±6	27±6	0.938
Body surface area (m ²)	1.9±0.3	1.9±0.2	1.8±0.3	0.030
Age at TOF repair (years)	5 (3–10)	8 (4–15)	4 (2–8)	0.002
Prior palliative shunt (%)	181 (38)	51 (52)	130 (36)	0.016
Comorbidities (%)				
Atrial fibrillation	118 (25)	60 (61)	58 (16)	<0.001
Atrial flutter/tachycardia	100 (21)	53 (54)	47 (13)	<0.001
Hypertension	125 (27)	45 (46)	80 (22)	<0.001
Hyperlipidaemia	193 (42)	60 (61)	133 (37)	<0.001
Coronary artery disease	57 (12)	24 (24)	33 (9)	<0.001
Current or prior smoker	93 (20)	31 (31)	62 (17)	0.005
Diabetes mellitus	72 (16)	20 (20)	52 (14)	0.253
Sleep apnoea	130 (28)	47 (47)	83 (23)	<0.001
Prior stroke	41 (9)	14 (14)	27 (8)	0.146
NYHA III/IV	79 (17)	24 (24)	55 (15)	0.061
Laboratory tests				
Haemoglobin (g/L)	141±25	140±18	141±28	0.628
Creatinine (mg/dL)	1.0±0.3	1.1±0.3	0.9±0.3	0.018
NT-proBNP (pg/mL)	255 (123–720)	1160 (133–2452)	210 (103–572)	0.126
Medications (%)				
Diuretics	84 (18)	29 (29)	55 (15)	0.003
Beta-blockers	106 (23)	33 (33)	73 (20)	0.012
Calcium channel blockers	59 (13)	24 (24)	35 (10)	0.001
RAAS antagonist	87 (19)	30 (30)	57 (16)	0.003
Warfarin	41 (9)	18 (18)	23 (6)	0.001
Direct oral anticoagulants	3 (0.7)	1 (1)	2 (0.6)	0.668

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	All (n=465)	CIED (n=99)	no CIED (n=366)	P values
Aspirin	116 (25)	37 (37)	79 (22)	0.005
Class I antiarrhythmic drugs	8 (2)	5 (5)	3 (0.8)	0.006
Class III antiarrhythmic drugs	35 (8)	19 (19)	16 (4)	<0.001
Digoxin	52 (11)	25 (24)	27 (8)	<0.001

CIED, cardiac implantable electronic device; NT-proBNP, N-terminal pro-B-type natriuretic peptide; NYHA, New York Heart Association; RAAS, renin angiotensin aldosterone system; TOF, tetralogy of Fallot.

Table 2

Haemodynamic data

Echocardiography	All (n=465)	CIED (n=99)	No CIED (n=366)	P values
Moderate RV enlargement * (%)	318 (68)	77 (77)	241 (67)	0.215
Moderate RV systolic dysfunction * (%)	137 (30)	49 (49)	88 (24)	<0.001
Moderate tricuspid regurgitation * (%)	92 (20)	37 (37)	55 (15)	<0.001
Moderate pulmonary regurgitation * (%)	324 (70)	69 (69)	254 (70)	0.390
Tricuspid regurgitation velocity (m/s)	3.1±0.7	3.1±0.7	3.1±0.7	0.652
Pulmonary valve peak velocity (m/s)	2.5±0.9	2.5±0.8	2.5±1.0	0.901
TAPSE (cm)	18±4	18±4	18±3	0.824
FAC (%)	40±10	36±12	40±10	0.051
RV S' (cm/s)	10±2	9±8	11±9	0.101
Medial E/e'	10±4	11±4	10±4	0.301
Lateral E/e'	7±3	8±3	7±3	0.178
LV ejection fraction (%)	58±8	55±10	58±8	0.002
LV mass index (g/m ²)	93±35	114±45	87±28	<0.001
Relative wall thickness	0.41±0.09	0.44±0.09	0.41±0.08	0.002
MRI	(n=164)			
RVEDV index (mL/m ²)	140±48	176±68	136±44	0.015
RVESV index (mL/m ²)	80±37	110±55	77±33	0.035
RV ejection fraction (%)	44±10	36±9	45±9	0.001
Catheterisation	(n=154)			
Right atrial pressure (mm Hg)	11±6	12±6	11±5	0.511
RVEDP (mm Hg)	14±5	15±5	14±6	0.655
Mean PA pressure (mm Hg)	24±9	24±9	23±9	0.123
LVEDP (mm Hg)	16±5	17±5	15±6	0.107
Mean arterial pressure (mm Hg)	88±13	90±13	88±13	0.448
Cardiac index (L/min*m ²)	2.3±0.6	2.2±0.5	2.4±0.7	0.039
PVR index (WU*m ²)	4.5±3.0	4.6±2.4	4.3±3.3	0.655
Cardiopulmonary exercise test	(n=181)			

Echocardiography	All (n=465)	CIED (n=99)	No CIED (n=366)	P values
Peak VO ₂ (mL/kg/min)	22±7	20±6	23±8	0.002
Peak VO ₂ (% predicted)	63±17	61±17	64±17	0.294
VE/VCO ₂ nadir	28±4	29±4	27±3	<0.001

* Quantitative assessment.

CIED, cardiac implantable electronic device; EDP, end-diastolic pressure; FAC, fractional area change; LV, left ventricle; PA, pulmonary artery; PAWP, pulmonary artery wedge pressure; PVR, pulmonary vascular resistance; RV, right ventricle; RVEDV, right ventricular end-diastolic volume; RVESV, right ventricular end-systolic volume; RVSP, right ventricular systolic pressure; TAPSE, tricuspid annular plane systolic excursion; VE/VCO₂, ventilatory equivalent for carbon dioxide; VO₂, oxygen consumption; WU*m², Wood units × metre squared. RVEDP: right ventricular end-diastolic pressure; LVEDP: left ventricular end-diastolic pressure.

Table 3**CIED data**

Pacemakers	n=41
Pacing site (%)	
Right atrium	2 (5)
Right ventricle	3 (7)
Dual chamber	35 (85)
Biventricular	1 (3)
Indications (%)	
Sinus node dysfunction	16 (39)
Heart block	26 (63)
Atrial arrhythmias/antitachycardia pacing	1 (3)
Cardiac resynchronisation therapy	1 (3)
Right atrial lead sensing threshold (mV)	1.9±0.9
Right atrial lead pacing threshold (V)	0.9±0.5
Right atrial lead impedance (Ω)	364±180
Right atrial pulse width (ms)	0.40±0.05
Right ventricular lead sensing threshold (mV)	8.3±5.9
Right ventricular lead pacing threshold (V)	1.2±1.0
Right ventricular lead impedance (Ω)	387±204
Right ventricular pulse width (ms)	0.40±0.05
Initial device implantation at Mayo Clinic	8 (20%)
Defibrillators	n=73
Indications (%)	
Aborted sudden cardiac death	3 (4)
Sustained ventricular tachycardia	43 (59)
Inducible ventricular tachycardia	16 (22)
Left ventricular ejection fraction <35%	12 (16)
Syncope	12 (16)
QRS duration >180 ms	3 (4)
Right atrial lead sensing threshold (mV)	2.0±0.9
Right atrial lead pacing threshold (V)	1.0±0.4
Right atrial lead impedance (Ω)	371±104
Right ventricular lead sensing threshold (mV)	9.1±4.4
Right ventricular lead pacing threshold (V)	1.3±1.1
Right ventricular lead impedance (Ω)	401±277
Right ventricular pulse width (ms)	0.60±0.10
Initial device implantation at Mayo Clinic (%)	9 (12)

CIED, cardiac implantable electronic device.

Table 4**Risk factors for defibrillator shock**

	HR (95% CI)	P values
Appropriate shock		
Age at time of device implantation (year)	1.32 (0.84 to 2.11)	0.324
Age at time of TOF repair (year)	1.21 (0.73 to 2.44)	0.187
Prior palliative shunt	1.04 (0.87 to 1.33)	0.155
Ventriculotomy incision	1.80 (0.62 to 5.86)	0.294
Left ventricular end-diastolic pressure >12 mm Hg	2.58 (0.91 to 4.04)	0.062
Mean pulmonary artery pressure >30 mm Hg	2.04 (0.51 to 5.56)	0.341
Right ventricular systolic pressure <50 mm Hg	1.71 (0.58 to 2.76)	0.381
Inducible ventricular tachycardia	1.91 (1.01 to 4.14)	0.062
Sustained ventricular tachycardia	1.95 (0.97 to 1.66)	0.068
QRS duration >180 ms	1.12 (0.67 to 2.02)	0.261
Class I antiarrhythmic drug	1.22 (0.74 to 2.43)	0.243
Class III antiarrhythmic drug	0.14 (0.04 to 0.54)	0.004
Inappropriate shock		
Age at time of device implantation (year)	1.54 (0.64 to 2.65)	0.121
Age at time of TOF repair (year)	1.26 (0.79 to 2.37)	0.143
Prior palliative shunt	1.21 (0.80 to 1.25)	0.143
Ventriculotomy incision	1.91 (0.54 to 5.11)	0.216
Left ventricular end-diastolic pressure >12 mm Hg	2.24 (0.90 to 3.96)	0.057
Mean pulmonary artery pressure >30 mm Hg	1.71 (0.29 to 4.07)	0.345
Right ventricular systolic pressure >50 mm Hg	1.88 (0.71 to 2.59)	0.308
Inducible ventricular tachycardia	0.93 (0.215 to 4.16)	0.654
Sustained ventricular tachycardia	1.93 (0.93 to 1.61)	0.063
QRS duration >180 ms	1.28 (0.56 to 2.55)	0.122
Class I antiarrhythmic drug	1.06 (0.32 to 4.41)	0.433
Class III antiarrhythmic drug	0.39 (0.12 to 0.72)	0.008

The incremental risks for continuous variables are expressed per unit increase. TOF, tetralogy of Fallot.