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Implantable cardioverter defibrillator therapy in paediatric practice: a single-centre UK experience with focus on subcutaneous defibrillation

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Aims Sudden cardiac death (SCD) risk can be managed by implantable cardioverter defibrillators (ICD). Defibrillation shocks can be delivered via ICD generator and/or intracardiac or subcutaneous coil configurations. We present our single-centre use of childhood ICDs.

Methods and results Twenty-three patients had ICD implantation, with median age and weight of 12.96 years and 41.35 kg. Indications included eight long QT; four hypertrophic cardiomyopathy; three Brugada syndrome; two idiopathic ventricular fibrillation; two post-congenital heart repair; two family history of SCD with abnormal repolarization; one catecholaminergic polymorphic ventricular tachycardia; and one left ventricle non-compaction. Twelve had out of hospital cardiac arrests prior to implantation. Techniques included 13 conventional ICD implants (pre-pectoral device with endocardial leads), 7 with subcutaneous defibrillation coils (sensing via epicardial or endocardial leads tunnelled to the ICD), and 3 with exclusive subcutaneous ICD (sensing and defibrillation via the same subcutaneous lead). Satisfactory defibrillation efficacy and ventricular arrhythmia sensing was confirmed at implantation. Follow-up ranged from 0.17 to 11.08 years. One child died with the ICD *in situ*. Ten children received appropriate shocks; five on more than one occasion. Five received inappropriate shocks (for inappropriate recognition of sinus tachycardia or supraventricular tachycardia). Five children underwent six further interventions; all had intracardiac leads.

Conclusion Innovative shock delivery systems can be used in children requiring an ICD. The insertion technique and device used need to accommodate the age and weight of the child, and concomitant need for pacing therapy. We have demonstrated effective defibrillation with shocks delivered via configurations employing subcutaneous coils in children.

Keywords Paediatrics • Implantable cardioverter defibrillators • Sudden death • Heart arrest • Electrophysiology

Introduction

Optimal management of sudden cardiac death (SCD) risk in paediatric practice is challenging. The clinical utility and benefit of implantable cardioverter defibrillator (ICD) therapy is well recognized, sometimes with adjunctive anti-arrhythmic medication, pacing manoeuvres, and sympathectomy.[1–5]

In adult practice, the standard of care for ICD therapy is pre-pectoral implantation of an ICD generator, connected to an endocardial pace-sense/defibrillation lead passed through the venous system to the right ventricle (RV) (with concomitant atrial and coronary sinus leads in dual chamber and cardiac resynchronization devices, respectively). An additional defibrillation coil may be situated in the superior vena cava, incorporated into the endocardial

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What's New?

- ICD implantation in childhood needs to be modified to the stage of the development of the child; conventional endovascular system, entirely subcutaneous and a hybrid approach are possible.
- Subcutaneous ICD devices can prove effective in children, and allow preservation of the vasculature and thus reduce lead related complications; a frequent problem in childhood.

lead, or as a separate coil. In other circumstances, a separate subcutaneous array can be implanted to improve defibrillation thresholds.

However, a recent development has been the entirely subcutaneous system. Subcutaneous-implantable cardioverter defibrillator (S-ICD; Cameron Health) has no endocardial leads and relies on a single subcutaneous lead (consisting of two sensing electrodes and a defibrillation coil) and customized arrhythmia discrimination.[6–8]

In paediatric practice, anatomical constraints militate against use of technology designed for adult practice. Furthermore, there is an established higher rate of inappropriate shock therapy and system failure (through lead fractures) in the paediatric population compared with the adult population.[9–11]

For these reasons, when paediatric ICD therapy is mandated by a high SCD risk, it is necessary to adopt novel approaches to ICD therapy delivery. We describe a clinical approach to paediatric ICD therapy with particular focus on the placement of defibrillation coils; subcutaneous, transvenous, and a novel hybrid approach of the two techniques. We conclude with our suggestions for ICD implantation based on the stage of growth of the child.

Methods

All patients who had an ICD device inserted under the age of 16 years in our institution (University Hospital Southampton NHS Foundation Trust, UK) from 1 January 2000 until the 31 December 2011 were identified using the internal hospital database.

Retrospective analysis of all patient notes, clinical letters and follow-up defibrillator assessments, and device interrogations were undertaken.

Pre-implantation data were collected to include the child's demographics (gender, age, and weight), underlying diagnosis (anatomical and electrical), the occurrence of pre-ICD cardiac arrest and pre-ICD management followed by the indication for ICD device implantation (primary or secondary prevention). Data at the time of implantation were collected to include implantation technique, types and arrangement of the ICD generator and leads, ICD programming details, and defibrillation efficacy, threshold and safety margin, and short-term complications within 28 days of implantation. Follow-up data were collected for each case including any long-term complications (after 28 days of implantation) and the number of defibrillation experiences the child underwent (appropriate and inappropriate). All the children were followed up at our institution until the age of 16, after which some cases transitioned care to their local hospital under the adult services. Local centres that were providing follow-up elsewhere were contacted to obtain information about shock therapy delivered during follow-up.

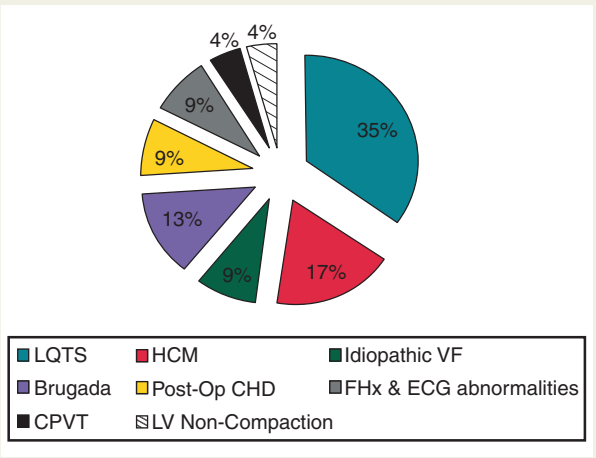

Figure 1 Pie chart demonstrating the ICD implant indications.

Twenty-three patients were identified; all of these children are included in this report with the maximum data sets we could obtain from electronic and paper records.

Results

Demographics

Twenty-three patients (age at implantation 0.92–16.00 years; median 12.96 years) had ICD implantation in the study period.

The median weight at implantation was 41.35 kg (range 7.30–82.00 kg).

Fourteen (61%) were male.

Implant indication

There was a range of indications for ICD insertion with congenital long QT syndrome being the commonest (8 cases; 35%).

Figure 1 displays the indications for ICD insertion.

Prior to ICD insertion, 12 of the 23 children (52%) had been successfully resuscitated from an out of hospital (OOH) cardiac arrest and had ICD insertion for secondary prevention.

The 11 children who underwent ICD implantation without an OOH cardiac arrest for primary prevention included: 4 children with congenital long QT with recurrent syncope despite maximal medical therapy; 3 children with Brugada syndrome (diagnosed with Ajmaline challenge in 2 and genetic positive in 1) all of them had a strong family history of SCD; 2 children (both siblings) who had non-specific repolarization abnormalities on the electrocardiogram (ECG) with episodes of syncope along with a strong family history of SCD with the mother having an ICD *in situ*; 1 child with hypertrophic cardiomyopathy (HCM) and a strong family history of sudden death (with a parent having an ICD *in situ*), and finally 1 child with HCM in a child with Noonan's syndrome and recurrent atrial arrhythmias.

Implant procedure

Implantable cardioverter defibrillator implantation techniques included 13 children with conventional ICD (pre-pectoral device

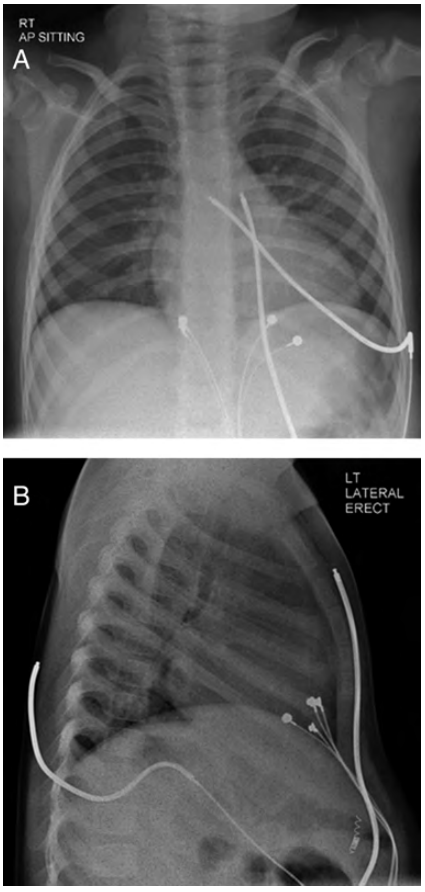

Figure 2 (*A* and *B*) Chest radiographs (AP and lateral) demonstrating the hybrid approach of a subcutaneous ICD coil along with epicardial leads for pacing in a 7.3 kg patient. The ICD generator is located in the abdominal tissues, due to size constraints of the child. A second subcutaneous coil is placed posteriorly to allow defibrillation to occur anterior to posterior.

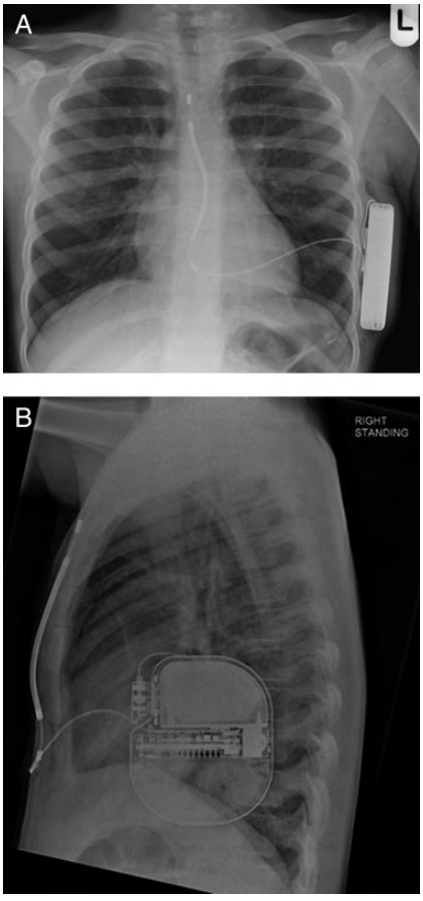

Figure 3 (*A* and *B*) Chest radiographs (AP and lateral) demonstrating the entirely subcutaneous ICD generator and coil in a 31 kg patient.

with endocardial leads). Seven children had ICDs with subcutaneous defibrillation coils (Medtronic 6996SQ coil). The 25 cm coil, approved for humanitarian use after achieving ethical and regulatory approval, is positioned subcutaneously, in an anterior and posterior position. Sensing is via epicardial leads placed during surgery or endocardial leads tunnelled to the ICD (*Figure 2*). Three children had exclusive subcutaneous ICD insertion with S-ICD (*Figure 3*). The age range of these children at time of the implantation was from 9.25 to 15.00 years (median 10.33 years) with a weight range from 31.38 to 54.31 kg (median 32.10 kg). Two of these children had long QT with OOH cardiac arrest. Since the S-ICD cannot deliver long-term pacing, it was used in conjunction with a conventional bipolar pacemaker to deliver the atrial pacing (AAI) necessary for one of these cases. One child had Brugada syndrome and a strong family history of SCD, but had remained asymptomatic.

In these three patients, pre-implant manoeuvres were undertaken to assess the ability of the Cameron Health discrimination algorithm to appropriately sense ventricular activity. No patients were screened out before implant. This screening process is undertaken with Cameron Health, and involves analysis of a standard 12-lead ECG and additional surface electrodes positioned to approximate the proposed subcutaneous electrodes. R- and T-wave amplitudes must fall within satisfactory limits to ensure effectiveness of the algorithm. In addition, exercise testing can be undertaken to examine morphology changes. If required, the same process is undertaken after permanent pacemaker implant, with testing at maximum bipolar output (unipolar pacing is incompatible). In addition, the upper pacing rate limit must be programmed below the ventricular arrhythmia zone. This approach has been used at our centre in the adult population requiring S-ICD with pre-existing pacing.

Defibrillation assessment and programming of implantable cardioverter defibrillator

Satisfactory defibrillation was confirmed at time of implantation in all children by the induction and termination of ventricular fibrillation (VF) with a 10 J safety margin, repeated twice as a minimum.

Table 1 Patient characteristics included in this study

	Gender	Diagnosis	OOH cardiac arrest?	Age at ICD insertion/ years	Weight at ICD insertion/kg	Implant radiology data		Lead and coil	
						Screening time/mins	Exposure dose/ microGymm^{-2}	Sensing lead	Defibrillation coil
Conventional ICD (Endocardial leads)									
1	M	Idiopathic VF arrest, previously well	Yes	15.25	60.00	6.2	302	Endocardial	Endocardial
2	M	Long QT syndrome Recurrent syncope despite maximal treatment	No	13.00	58.90	1.18	27	Endocardial	Endocardial
3	M	Brugada syndrome, family history of sudden death	No	12.92	41.35	u/a	u/a	Endocardial	Endocardial
4	M	OOH VF arrest, no cause found but history of recurrent syncope	Yes	15.33	82.00	10.44	561	Endocardial	Endocardial
5	F	Abnormal repolarization changes on ECG, Mum has ICD *in situ* for strong FHx of sudden death	No	13.92	47.50	u/a	u/a	Endocardial	Endocardial
6	F	Abnormal repolarization changes on ECG, Mum has ICD *in situ* for strong FHx of sudden death	No	9.67	31.12	0.08	3.3	Endocardial	Endocardial
7	M	Long QT syndrome Recurrent syncope despite maximal treatment	No	13.92	58.90	u/a	u/a	Endocardial	Endocardial
8	M	HOCM, Noonan's syndrome, Recurrent atrial arrhythmias and syncope	No	16.00	56.00	2.35	173	Endocardial	Endocardial
9	F	HCM, family history of sudden cardiac death, Mum has ICD *in situ*	No	15.75	58.20	u/a	u/a	Endocardial	Endocardial
10	M	HCM	Yes	10.08	30.89	9.29	177.2	Endocardial	Endocardial
11	F	HOCM	Yes	8.42	23.80	4.13	162	Endocardial	Endocardial
12	F	Long QT Syndrome Recurrent syncope despite maximal treatment	No	15.33	59.15	3.5	119	Endocardial	Endocardial
13	M	Long QT syndrome	Yes	14.67	68.90	15.29	464	Endocardial	Endocardial
Subcutaneous defibrillation coils with sensing via epicardial or endocardial leads (*Figure 2*)									
14	M	Brugada syndrome, family history of sudden death	No	11.50	35.81	0.52	u/a	Endocardial	S/C
15	F	Post-op LV histiocytoma and MV repair	Yes x 2	0.92	9.10	u/a	u/a	Epicardial	S/C
16	F	LV non-compaction and intraventricular conduction delay	Yes	1.08	7.30	1.11	3.8	Epicardial	S/C
17	M	Catecholamine polymorphic VT	Yes	1.25	9.30	0.15	0.8	Epicardial	S/C
18	M	Previous DORV and sub-aortic stenosis repair, with MV replacement	Yes x 2	15.83	69.20	0.2	12.1	Endocardial	S/C
19	M	Long QT and dilated cardiomyopathy	Yes	7.92	24.60	u/a	u/a	Endocardial	S/C
20	F	Long QT syndrome Recurrent syncope despite maximal treatment, FHx of sudden death	No	10.92	37.20	1.02	u/a	Endocardial	S/C
Exclusive subcutaneous ICD (Cameron Health Device) (*Figure 3*)									
21	M	Brugada syndrome	No	9.25	32.10	0.1	5	S/C	S/C
22	F	Long QT syndrome	Yes	15.00	54.31	u/a	64.8	S/C	S/C
23	M	Long QT syndrome	Yes	10.33	31.40	0.02	6	S/C	S/C

	Programming data								Follow-up period		
	VF rate / b.p.m.	VT rate / b.p.m.	Initial shock/J	Max. shock energy/J	Shock vector	Detection	Redetection	ATP	Duration, years	Appropriate defibrillation?	Key events
Conventional ICD (Endocardial leads)											
1	222.2	250	35	35	RV to generator	18/24	12/16	X10	5.08	Not needed	Inappropriate defibrillation
2	222.2	No	35	35	RV to generator	18/24	12/16		5.17	Not needed	None
3	250	200	35	35	RV to generator	u/a	u/a		9.92	Not needed	None
4	200	230.7	35	35	RV to generator	30/40	12/16	X5	1.83	Yes	Inappropriate defibrillation
5	222.2	No	35	35	RV to generator	18/24	12/16		4.92	Not needed	None
6	222.2	No	35	35	RV to generator	18/24	12/16		4.92	Not needed	None
7	222.2	No	35	35	RV to generator	18/24	12/16		4.33	Not needed	None
8	200	No	25	35	RV to generator	30/40	18/24		7.50	Not needed	Inappropriate defibrillation
9	214.2	250	35	35	RV to generator	30/40	12/16	X10	6.00	Not needed	Lead migration
10	187.5	240	35	35	RV to generator	18/24	12/16	X5	10.83	Yes	Possible superficial wound infection
11	150	250	35	35	RV to generator	18/24	12/16	u/a	1.17	Yes	Died (shock refractory VF)
12	u/a	u/a	u/a	u/a	RV to generator	u/a	u/a	u/a	5.92	Not needed	None
13	200	No	35	35	RV to generator	18/24	12/16		2.42	Yes	None
Subcutaneous defibrillation coils with sensing via epicardial or endocardial leads (*Figure 2*)											
14	u/a	No	21	31	u/a	u/a	u/a		6.17	Not needed	None
15	200	240	35	35	A-P coil	18/24	12/16	During charging only	8.33	Yes	Epicardial lead migration then lead outgrown
16	200	No	18	35	A-P coil	18/24	12/16		1.83	Yes	Inappropriate defibrillation
17	200	240	35	35	A-P coil	24/32	12/16	X3	1.75	Yes	None
18	200	250	35	35	A-P Coil	30/40	12/16	X3	4.42	Yes	Infective Endocarditis
19	165	No	31	31	A-P Coil	u/a	u/a		6.33	Not needed	Lead outgrown
20	222.2	No	35	35	RV to generator	18/24	12/16		4.92	Yes	Inappropriate defibrillation
Exclusive subcutaneous ICD (Cameron Health Device) (*Figure 3*)											
21	240	No	80	80					1.08	Not needed	None
22	240	No	80	80					1.33	Not needed	None
23	240	No	80	80					0.42	Not needed	None

OOH, out of hospital; HCM, hypertrophic cardiomyopathy; HCOM, hypertrophic obstructive cardiomyopathy; VF, ventricular fibrillation; VT, ventricular tachycardia; Post-op, post-surgical correction; DORV, double outlet right ventricle; FHx, family history; CPVT, catecholaminergic polymorphic ventricular tachycardia; LV, left ventricle; RV, right ventricle; MV, mitral valve; S/C, subcutaneous; u/a, unavailable.

Where applicable, AAI pacing was continued during testing. We had no cases of unsuccessful defibrillation at the time of testing in this cohort.

Configuration and programming data were available for 18 patients, with only limited data available in other cases where follow-up had continued at another centre. The S-ICD devices select the best of three sensing vectors, and has a unique arrhythmia detection algorithm based on rate detection and feature extraction. It delivers a non-programmable shock of 80 J using with an automated vector capable of reversing polarity in the event of failure to cardiovert.

For the devices with subcutaneous coils, the shock vector is from the anterior cathode to posterior anode, with the generator inactive in the circuit. This set up was chosen since it ensures the ongoing safety of the shock vector in the face of growth and migration, especially where an abdominal generator site is chosen. Of the other devices, one was set to deliver a shock from generator to RV coil. All other devices were set to deliver a shock from RV coil to generator. Three devices delivered an initial shock at below maximum output, with all further shocks at maximum. All other devices delivered all shocks at maximum output. The shock vector was reversed on the final sequence in all cases.

Detection was set to 18/24 in 10 cases, and 30/40 in four cases. One device was set to 24/32. Redetection was appropriately reduced in all cases. Sixteen devices had a VF detection zone set at or above rates of 200 b.p.m. The highest zone was set at 250 b.p.m. in a device and 240 b.p.m. in the Cameron Health devices. The lowest detection rate was set at 150 b.p.m. Nine devices had a separate ventricular tachycardia zone, all with rates $>$200 b.p.m.

Procedural safety

No procedural complications occurred during implantation. There were no device infections or revisions. One child had a post-operative area of erythema over the wound that was treated with a 7-day course of oral antibiotics, and resolved with no long-term complications.

Follow-up

Overall follow-up for all types of ICD ranged from 0.17 to 11.08 years (median 4.92 years). Nine children (41%) received appropriate defibrillation shocks for ventricular fibrillation (five children on more than one occasion during follow-up).

Five children (22%) received inappropriate shocks, all due to inappropriate recognition and discrimination of sinus or supraventricular tachycardia. All of these children had either endocardial or epicardial sensing leads. Further manoeuvres were employed to rectify this with software re-configuration to extend the number of intervals of tachycardia that needed to be detected to declare the commencement of an episode of ventricular arrhythmia, that satisfied device detection criteria (to 120/140 beats) (Medtronic^{TM} Inc.).

One child died with the ICD *in situ*. This child had shock-refractory ventricular fibrillation with HCM. The ICD did defibrillate multiple episodes appropriately, but arrhythmia re-initiation lead to therapy exhaustion in this case.

Five children underwent six additional interventions during the follow-up period. All of these children had an intracardiac lead (three children had outgrown the leads, one lead fracture, one lead migration, and one child had late development of infective endocarditis of an underlying congenital heart lesion necessitating the removal of the intracardiac ICD leads, 2 years after insertion).

No complications occurred in the children with the exclusive subcutaneous ICD device over a median follow-up period of 0.83 years (range 0.17–1.08 years).

Table 1 displays the details of the 23 children in this paper. All children until the age of 16 years had routine yearly chest X-rays, six monthly ICD interrogations and clinical review.

Anti-arrhythmic medication

Twelve patients (52%) had at least one anti-arrhythmic medication prescribed for at least 1 month prior to ICD implantation. Three children were prescribed two medications. No children had more than two anti-arrhythmic drugs prior to ICD implantation. Of these 12 patients, 11 received β-adrenoreceptor blockers, three received Amiodarone and one child received Flecainide. Following ICD implantation 15 children received anti-arrhythmic medication (in all cases this was a β-adrenoreceptor blocker).

Discussion

In adult practice, ICD therapy is supported by a substantial evidence base for management of the risk of SCD.[12,13] There is increasing uptake of ICD implantation in children, with data that demonstrates safety and efficacy in the paediatric population.[11,14,15] However, as ICD therapy in childhood is a relatively recent development with technology improvement over the past decade, little data exists for very long-term outcomes. Our data suggests that these children do very well with ICD devices *in situ*, with a median follow-up of 4.9 years with over 65% of children followed up for more than 5 years. This long-term follow-up is important, as over a 5 year period many children will have a growth spurt to some degree, which raises issues relating to the risk of lead migrations and revisions. We have demonstrated the ability to manage these children safely and effectively, but it is important to take into account many technical factors when considering paediatric ICD implantation.

The sensing of ventricular electrophysiology can be via endocardial, epicardial, or subcutaneous sensing leads, although work is ongoing into other methods such as intrapericardial devices.[16] Implantable cardioverter defibrillator shock therapy delivery has traditionally been via intracardiac defibrillating coils, but can also be delivered by subcutaneous coils. A transvenous inserted endocardial ICD lead is the established approach in adults, with an insertion technique that is similar to that for endocardial pacing. Problems exist with this technique in the paediatric arena. Many papers consider the difficulties of pacing in children.[10,17] Many of these difficulties apply to pediatric ICD usage with additional risks related to appropriate placement of endocardial coils in the paediatric heart.

Thus, the transvenous approach in children shares similar risks to that with adults; pneumothorax, endocarditis risk, and thromboembolic phenomena. However, the child has some unique problems including difficult vascular access, need for linear growth, narrower vessel lumens with higher risks of thrombosis/fibrosis,

and higher lead failure rates. Lead failure is more common as the child grows.[10] Furthermore, an endocardial ICD lead inserted in childhood will almost certainly require a lead revision in later life—a procedure with significant risks. In our cohort, 38.5% of the transvenous systems needed revision. Adult data suggest revision rates of 12–20% for a similar timeframe as our cohort.[18–20]

Until recently, use of defibrillation coil(s) mounted on a transvenous lead coupled to an ICD generator, placed in a pre- or post-pectoral pouch was the principal defibrillation configuration employed by ICDs in clinical paediatric practice. However, due its complexity of build, the ICD pace-sense/defibrillation coil is more vulnerable to physical damage and lead break than the simpler but more robust pacing leads. The high ICD lead failure rate is well established across all manufacturers.[21,22] Failure results in lead extraction and system revision which itself is associated with significant morbidity and risk.[23]

The subcutaneous ICD device from Cameron Health is an entirely subcutaneous sensing and defibrillating ICD. It would appear to have some advantages when considered for children. It does not require the insertion of any leads into the vasculature of the child, leaving the option of transvenous ICD insertion in later life. Increased inappropriate shock rate has been reported by one centre,[24] but this may reflect relative inexperience with patient selection as this has not been borne out in other reports.[25] Intuitively, it is arguable that it is easier to replace a subcutaneous lead compared to an endocardial lead system, although clinical trial or registry data does not yet exist to support this assertion. Furthermore, the current commercially available subcutaneous device generator is large and may not be implanted comfortably in smaller children. In the children we have implanted this system in, we have had no reported concerns regarding the aesthetic appearance or discomfort with the relatively larger devices. An entirely subcutaneous ICD system does not require a sternotomy for implantation, which is required when (for reasons of disease or anatomy) epicardial pace-sense leads must be placed for conventional ICD therapy application. However, in this scenario an endocardial bipolar lead can be placed and tunnelled to the ICD generator which is then attached to subcutaneous defibrillation coils. Some adult data suggests that subcutaneous ICDs are equally as effective as the established transvenous ICDs.[6] In our cohort, none of our patients with exclusive subcutaneous ICD systems required defibrillation. Therefore, we are unable to comment on its shock efficacy based on our data. A further limitation of this system is inability to provide anti-bradycardia or tachycardia pacing, as no myocardial electrical interface is present for the delivery of endocardial or epicardial pacing.

To our knowledge, limited literature exists regarding the use of exclusive subcutaneous ICDs in children. The data that does exist in childhood often involves epicardial leads or transvenous endocardial ICD use, and usually in children over 30 kg in weight.[7]

We have utilized a variety of approaches for application of ICD therapy including an entirely subcutaneous approach and a hybrid approach combining endocardial or epicardial pace-sense leads with subcutaneous defibrillation coils.

The approach adopted has been determined by anatomical considerations, therapy need, and patient preference. It has been our intention to minimize invasion of the vasculature, as we are conscious that ICD therapy is likely to be required lifelong in these patients and will need to be re-deployed in other formats in early adult life. By preserving the vasculature, we are protecting the options for future therapy and reducing the morbidity and mortality risks associated with early endocardial ICD lead failure. Furthermore, it is our experience that subcutaneous defibrillation shock configuration is more forgiving of growth-related change than is perhaps the case with endocardial leads where adhesion to intravascular structures limits spontaneous adaptation to the changing anatomy of the growing child. Our limiting factor when considering subcutaneous ICD insertion in children is the relative size of the child to the device.

Limitations

By the nature of the low implantation rates of ICD in children across many centres, our sample size is small although no children were excluded. We furthermore acknowledge that this is a single-centre experience, limited to a fixed number of paediatric ICD implanters. Due to a change in our database system during the review period, some data were unable to be obtained from the time of implantation despite extensive attempts. It is important to appreciate that the number of Cameron Health patients is particularly small (although this is a new technology). Within this group, no children required appropriate defibrillation. We did not conduct patient interviews as part of this follow-up period, hence are unable to comment on the quality of life impact that ICD implantation had. It is our experience that once a child has had an OOH cardiac arrest, or is at high risk, many parents are unhappy to be discharged without an ICD *in situ*. Further work has considered the psychological impact and quality of life in those with ICD in more detail.[26,27]

Conclusions

We have reported a variety of approaches to the use of ICD therapy in paediatric and congenital heart disease practice, which have been intended to meet the particular challenges of altered cardiac anatomy or patient size. These have included hybrid subcutaneous defibrillation and endocardial/epicardial pacing leads, entirely subcutaneous defibrillation and bespoke ICD detection features to allow very long periods of ventricular tachycardia to self-terminate and so avoid unnecessary shock therapy. We suggest that a systematic approach should be employed to assess optimal provision of protection from SCD at different stages during early life. Considerations include the need to preserve the vasculature from ICD lead-related damage, effective arrhythmia sensing and discrimination, and efficacy of defibrillation therapy. Thus we hypothesize that (given current commercially available technologies) the approach should be:

- *In infancy:* subcutaneous defibrillation but endocardial (only V lead) or epicardial sensing (A and V leads particularly when pacing required and if access to the epicardium for lead implantation has not been compromised by previous surgery). This will require re-assessment of system efficacy at intervals.

- *In childhood:* entirely (arrhythmia sensing and no conventional pacing) subcutaneous defibrillation if the child is large enough for current technologies, or a 'hybrid' subcutaneous ICD system with separate pacemaker leads to epicardium or endocardium or an entirely subcutaneous (arrhythmia sensing and no conventional pacing) ICD and separate pacemaker (if pacing to treat bradycardia is indicated). Further work is needed to consider the feasibility of hybrid approach.
- *Post-pubertal growth spurt:* if pacing required: a conventional ICD or if no pacing required: an entirely subcutaneous (arrhythmia sensing and no conventional pacing) defibrillator or a subcutaneous ICD (arrhythmia sensing and no conventional pacing) and separate pacemaker.

We have demonstrated the efficacy of these approaches in a small case series with long follow-up.

Conflict of interest: M.J.G., J.G., and M.H. have no conflict of interests to declare. J.M.M. receives research support, speaker bureau honoraria, and consulting fees from Medtronic Inc., St Jude Inc., Boston Scientific Inc., and Cameron health Inc. J.A.R. receives an unrestricted educational grant from Medtronic.

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