

REVIEW ARTICLE

Implantable Cardioverter-Defibrillators in Children: Innovation to Design a Pediatric Implantable Cardioverter-Defibrillator

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ABSTRACT. Although the implantable cardioverter-defibrillator (ICD) is now established as safe and effective for prevention of sudden cardiac death (SCD) in children,^{1–3} pediatrics accounts for less than 1% of market share, inhibiting corporate financial incentive for development of a pediatric-specific device. Over the past two decades since the ICD became clinically available, progressive downsizing of devices and leads has allowed for their use in children. However, the smallest of children and those with particular forms of congenital heart disease (CHD) often require customized implant techniques. Children have different implant indications, implant techniques, programming issues, psychosocial impact, and follow-up care considerations. Compared with the typical adult ICD patient, most pediatric ICD patients outlive their device by decades. This necessitates proper advance planning to maximize device and lead longevity, vascular patency, and other chronic management issues. This paper will briefly review the specific indications and implantation issues unique to pediatrics and will suggest future innovations to consider for developing a pediatric defibrillator.

KEYWORDS. implantable cardioverter-defibrillator, congenital heart disease, pediatrics, lead.

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Pediatric-specific indications for defibrillator therapy

Identifying and selecting appropriate pediatric implantable cardioverter-defibrillator (ICD) candidates involves considering the cumulative risks of sudden cardiac death (SCD) from the underlying disease substrate compared with the procedural risks of ICD implantation and chronic management issues. This involves an analysis of the competing risks of malignant arrhythmia with other causes of mortality, balanced against the complications from ICD therapy, including surgical morbidity, inappropriate shocks, and multiple system revisions. Recommendations for secondary prevention in children are similar to ICD implantation guidelines developed for adults,⁴ despite a paucity of pediatric randomized controlled trials. Several retrospective studies demonstrate efficacy of ICD therapy in young patients.^{1,5–6}

Class I indications in pediatric patients include aborted SCD or hemodynamically significant sustained ventricular tachycardia without a reversible cause. There are less data on which to base guidelines for pediatric primary prevention. Expert consensus and extrapolation from adult studies have guided primary prevention recommendations based on Class IIb evidence levels.⁴ In general, ICD therapy may be considered in selected high-risk children with presumed or documented risk factors, such as family SCD history, non-invasive measurements (e.g. QTc, ventricular septal thickness, etc.), medication intolerance, or specific genetic mutations. Presymptomatic recognition of at-risk patients is ideal, as SCD may be the sentinel (and sometimes fatal) event in a previously asymptomatic child. Extrapolation from adult randomized clinical trials, a limited number of retrospective pediatric studies, and expert consensus are only a surrogate for well-designed prospective pediatric clinical ICD trials. There are clinical scenarios in pediatrics that do not fall under the typical rubric of published guidelines. For example, infants with malignant arrhythmias may benefit from ICD therapy but require more complex implantation procedures. Primary prevention ICD therapy in infants has not been systematically evaluated.

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Pediatric-specific ICD implantation

Implantation of ICDs in children and congenital heart disease (CHD) patients requires customized preprocedural planning, with individualized techniques for each patient. There are important considerations with regards to size, growth, and anticipated activity. Young patients are likely to outlive the current-generation devices and leads, necessitating multiple replacement and complex extraction procedures.

Pediatric ICD recipients were previously limited to receiving epicardial systems because of anthropometric issues, longitudinal growth, ICD size, and transvenous lead dimensions.⁷ Technological advances such as biphasic defibrillation waveform and active generators, combined with a progressive reduction of generator size and lead dimensions, allowed transvenous ICD systems for adolescent pediatric patients. Transvenous leads in small children carry risks of venous occlusion, infection and endocarditis, potential embolic events in the presence of an intracardiac shunt, and lead damage. Epicardial ICD systems are inherently a more invasive procedure, have a higher incidence of lead/patch failure, and a possibility of developing constrictive pericarditis with scarring from the relatively large defibrillation patches in relationship to smaller heart size. However, regardless of implant route, defibrillator lead failure rates remain unacceptably common in pediatric patients.

Implanting an ICD in a child necessitates the consideration of potential quality of life and emotional and psychosocial development issues.⁸⁻¹⁰ These issues ideally should be addressed prior to the initial implant in order to minimize long-term psychosocial impact.

Issues with using off-the-shelf ICD products designed for adults

The most obvious differences in needs for defibrillator systems in children relate to size. The relatively large generator size and lead diameter and length are challenges for implantation in smaller pediatric patients and also carry substantial concerns for chronic follow-up issues. Often, these size discrepancies force alternate procedural strategies, such as abdominal generator placement or non-transvenous lead placement, particularly in very small children or those with particular forms of CHD. Besides the size issues, there are important considerations regarding shock strength, programming issues, and overlap of tachycardia detection criteria with normal pediatric heart rates.

Pediatric-specific customized ICD implantation

Limitations with standard transvenous and epicardial ICD systems in children and CHD patients have prompted individual clinical investigators to develop novel implantation techniques. Animal models and computer modeling studies have shown the feasibility of functional ICD systems without the need for transvenous shocking coils or epicardial patches.

For example, subcutaneous array and coils were originally designed for adjunctive use in order to lower the defibrillation threshold. However, several independent groups almost simultaneously reported innovative use of subcutaneous arrays or coils for defibrillation in children.¹¹⁻¹³ These novel subcutaneous ICD systems negated the requirement for transvenous coils or epicardial patches, but still utilized epicardial leads for pacing and sensing (Figure 1, upper right). Subcutaneous ICD systems have now been reported in pediatric multicenter studies, with appropriate tachycardia detection and effective delivered therapies during intermediate-term follow-up.¹⁴ There are also completely leadless ICD systems currently in early clinical use and investigation.^{15,16} However, current generation leadless ICDs may not be suitable for small children, as the generators are relatively large in order to accommodate the significantly higher (at least twofold) defibrillation outputs necessary. Furthermore, the currently available totally subcutaneous ICD system does not have the capability for chronic antibradycardia pacing (other than emergent post-shock transcutaneous pacing) or antitachycardia pacing, which may be indicated in a substantially higher proportion of pediatric and congenital ICD recipients. However, the subcutaneous ICD could be potentially downsized to a smaller pediatric-specific device, as these patients may require lower defibrillation energy requirements and therefore need smaller capacitors and batteries. These exciting concepts will need to be tested in preclinical and pediatric clinical situations.

Non-standard configurations for pediatric implantable defibrillator systems have also been designed with the placement of leads in the pericardial or pleural space.^{17,18} This approach circumvents the need for epicardial patches or transvenous hardware while providing defibrillation capability. Various off-the-shelf transvenous ICD leads, superior vena cava leads, or subcutaneous coils have been used for this purpose, placed in the posterior pericardium (Figure 1, lower right). Additional pacing/sensing leads can be placed on the epicardium. This procedure may be performed via a less-invasive videoscopic technique along with a small subxiphoid incision.

Interestingly, when non-transvenous ICD systems were retrospectively evaluated, it turned out that they had poorer longevity than traditional transvenous ICD systems in children.¹⁹ Therefore, non-transvenous ICD systems are not ready to supplant standard ICDs in the majority of pediatric patients but still have an important role for those patients with limited alternatives. Non-standard systems might be used as the initial device until the child reaches a larger size and can potentially accommodate a transvenous standard ICD system.

Moving into the future: What modifications in the ICD pulse generator or lead would the pediatric electrophysiologist consider desirable?

ICD generator

Current generation ICD devices, although significantly smaller in volume than their predecessors, still require

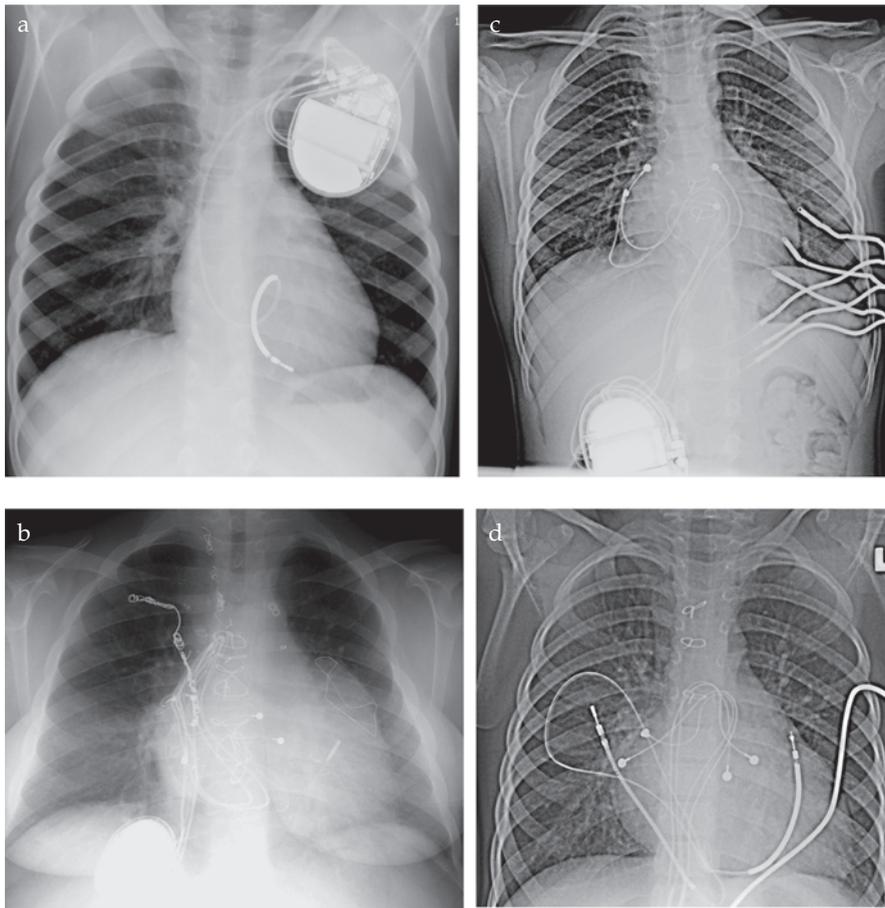


Figure 1: Examples of four routes of implantable cardioverter-defibrillator (ICD) implantation in children described in the text. The (a) is a transvenous single-coil ICD system, with the lead looped in the right ventricle in a small 27-kg 7-year-old child with hypertrophic cardiomyopathy and resuscitated from a ventricular fibrillation arrest. The (b) is an example of epicardial patch ICD system along with epicardial suture-on dual-chamber pace/sense leads in a young teenager with single ventricle and failing Fontan physiology. The (c) is an example of a subcutaneous array ICD system in a boy with malignant catecholaminergic polymorphic ventricular tachycardia, using epicardial dual-chamber suture-on leads for sensing and DDD pacing. The (d) is an example of a pericardial ICD lead system in a 3.5-year-old with congenital heart disease and dilated cardiomyopathy, using a transvenous-design ICD lead placed in the posterior pericardium, DDD epicardial pacing leads, as well as a subcutaneous coil in the left lateral chest wall. The lead that is seen in the right pleural space is the pace/sense portion of the ICD lead, which is capped since the epicardial pacing leads are used for sensing and pacing in this configuration. (Reprinted from reference 22, with permission from Elsevier.)

the implanting operator to produce significant tissue injury to implant the device. In infants and very small children, it still becomes a technical challenge to find suitable space for the generator. In small and/or thin children, its resultant cosmetic deformity impacts quality of life and may have secondary effects on the patient's self-image. For the smaller pediatric patients, reduction in ICD generator size will enable improved cosmetic results and reduce the potential for pocket erosion and infection. Many of the cardiac diseases that necessitate ICD implant in childhood (repaired CHDs, congenital long QT syndrome, and hypertrophic cardiomyopathy) are associated with repolarization disorders and "unusual" T waves. T-wave oversensing is a common problem in this group of patients, with few readily available options for handling the inappropriate sensing. Software or filter techniques that reduce device susceptibility to T-wave oversensing should be incorporated into the "pediatric-friendly" device. The higher max-

imum heart rate of the pediatric patient necessitates device manufacturers to allow safe and more intelligent programming of higher bradycardia pacing rates, pacing upper rate limits, sinus and supraventricular tachycardia discriminators, and ventricular tachycardia detection rates. Creative changes in the defibrillation waveform can allow for lower defibrillation thresholds and smaller devices.

ICD lead

Currently designed ICD leads, although relatively effective and robust in the adult patient, are known to have a high failure rate in the pediatric patient population, regardless of implant route. Pediatric patients require the design of a significantly different type of lead than is used for the adult ICD patient. The "pediatric-friendly" ICD lead should incorporate insula-

Table 1: Implantable defibrillator lead route options in pediatrics

	Advantages	Disadvantages
Transvenous	Relatively easy implant; common use; approved indication	Lead fractures; extractions difficult; vascular obstruction
Epicardial patch	Long history, follow-up; approved use; surgeons familiar; good DFT	Patch failure; buckling; constrictive pericardial physiology
Subcutaneous array or coil	No transvenous coil or epicardial patch; minimally invasive	Limited long-term data; higher DFT
Pericardial coil	No need for transvenous access or epicardial patch; low DFT	Requires surgeon; adhesions may limit VATS; limited follow-up data
Subcutaneous leadless ICD	No need for transvenous or epicardial access; minimally invasive	Limited long-term data; higher DFT; no chronic pacing or ATP; large can

DFT: defibrillation threshold; VATS: video-assisted thoracoscopic surgery; ATP: antitachycardia pacing; ICD: implantable cardioverter-defibrillator.

Note: The subcutaneous and pericardial techniques are not FDA-approved indications for these devices.

tion and conductor materials that allow for greater flex-induced distortions, since these leads are implanted in a more “active” environment, and these enhancements may also be of value for adult patients. Using engineering imagination, designing leads that stretch for anticipated linear growth and inhibit tissue fibrosis and binding would be welcome improvements for pediatric ICD recipients (and their implanters/physicians). These changes would be anticipated to improve ICD lead longevity in the pediatric patient. Shorter ICD lead lengths could also be beneficial by allowing a reduction in pocket bulk from redundant lead slack, having the potential to decrease pocket infection rates and improve cosmetic appearance. Incorporation of novel agents/materials in the ICD lead insulation that can prevent clot formation on the lead or fibrous band attachment to the venous tissue would enhance the ability to handle the inevitable need to extract the ICD lead in this patient population. Reduction in the tip to coil setback distance will allow the implantation of ICD leads in smaller and younger patients and will help avoid tricuspid valve injury secondary to the ICD coil resting on the tricuspid valve and potentially causing interference with normal valve function.

In summary, there are several methods from which to choose when implanting devices in pediatric patients. The advantages and disadvantages of current reported methods for ICD lead placement are summarized in **Table 1**. The risks and benefits of each approach need to be considered for each patient and disease type. Individualized therapy choices may need creative, customized, non-standard, or hybrid approaches. Although there are not specific weight or size cutoff criteria, a clinical practice of limiting placement of transvenous ICD leads in small children is reasonable due to concerns of venous patency and lead failure with patient growth, and potentially more difficulty with lead extraction.^{3,20,21} This strategy of using a non-transvenous lead for their initial system allows the protection of an implantable defibrillator in small children without limiting future vascular accessibility. The pediatric electrophysiology community has an imperative to provide the best means of achieving long-term safe and effective ICD therapy for children. Although progress in ICD technology has been truly remarkable over the past two decades, the current

limitations in pediatric patients should be an impetus for device manufacturers, biomedical engineers, researchers, and physicians to work together in designing optimal ICD systems for children. Although market forces may not be the driver for pediatric ICD research and development, the unique needs of this patient population, the potential life-years saved per patient, and corporate charitable goodwill, along with anticipated governmental regulations for pediatric devices, should hopefully be sufficient motivation for moving forward with designing a true pediatric ICD.

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