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Update on pacing and implantable cardioverter defibrillators in children

Anne M. Dubin *

Stanford University, Stanford, CA, USA

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ABSTRACT

Pacemakers and implantable cardioverter defibrillators are important therapies in the pediatric and congenital heart disease population. However, these therapies, while potentially life-saving, have major limitations which are particular to the pediatric age group. Lead malfunction, inappropriate discharges, and venous obstruction all complicate device usage in this group. This review will address issues with pacemaker and defibrillator use in the pediatric and congenital heart disease population and discuss potential advances in the area in the next 5 to 10 years.

1. Introduction

Innovations in pacemakers and implantable cardioverter defibrillator technology have largely been driven by the adult market. This is not surprising, as children and congenital heart disease patients make up less than 1% of the device population. Thus pediatric innovations in this area are limited to new and novel usage of existing therapies. While these pediatric innovations have allowed protection in the smallest of patients, they have also highlighted the many issues and complications the pediatric electrophysiologist will encounter when using these technologies.

2. Size and growth

With recent advances in pediatric congenital cardiothoracic surgery, even the smallest children with severe congenital heart disease are able to survive to adulthood. These children often exhibit serious sequellae of their disease, including arrhythmias and heart block. It is therefore not uncommon for newborns to require permanent pacemaker therapy, and pacemakers have been implanted into children as small as 1000 g [1]. Life threatening arrhythmias have prompted pediatric electrophysiologists to push the boundaries of ICD therapy to protect even the smallest of patients. Stephenson and colleagues reported the multicenter experience with "novel" ICD placement which usually involved a subcutaneous, pericardial or pleural ICD coil and an epicardial sensing lead [2] (Fig. 1). These novel techniques have allowed implantation of an ICD in children as young as a few months of age.

It is common to implant devices epicardially (or, as mentioned above, in the pericardial or subcutaneous space in ICDs) in infants and small children, with a transition to a transvenous system in late childhood and adolescence. There are advantages and disadvantages to both approaches. An epicardial system spares the venous anatomy, avoiding venous thrombosis and the need for future lead extractions, but may require a mini-sternotomy, and historically has a higher lead failure rate than an endocardial system [3].

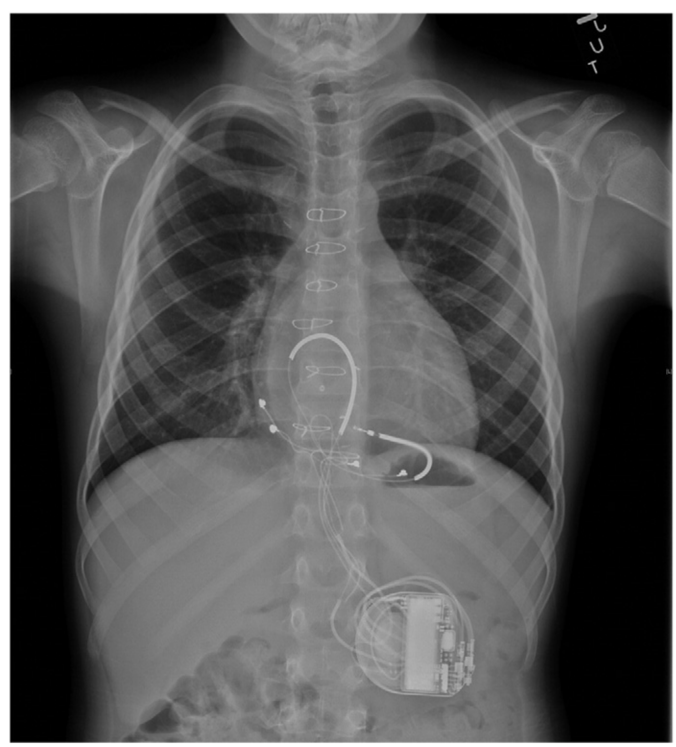

Fig. 1. Novel ICD lead implantation, ICD coil placed in pericardial space with pace/sense lead placed on ventricle.

* 750 Welch Rd., Suite 325, Palo Alto, CA 94303, USA. Tel.: +1 650 723 7913; fax: +1 650 725 8343.
E-mail address: amdubin@stanford.edu.

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Pediatric practitioners need to be fully cognizant of the fact that pediatric patients have a higher potential for growth, activity and longevity. This influences type of device and lead systems utilized. A child has the need for higher basal heart rate and thus will deplete batteries faster than an adult will. Unfortunately a large battery tends to translate into a large device, which can cause issues with erosion. Thus the pediatric electrophysiologist needs to balance the benefit of fewer operations to the risk of pacemaker erosion when contemplating which device to use.

Children will grow almost four-fold from infancy to adulthood with major growth spurts seen in early childhood and adolescence. Children also tend to be more active than adults and put greater stress on device hardware. These devices need to be fully functional for 70–80 years as opposed to the 20–30 year time frame for adult patients. These factors all lead to a higher lead failure rate in children than adults, with lead dislodgment and break more commonly seen [4] (Fig. 2). These lead failures result in either lead abandonment with a risk of venous obstruction and lead interactions, or the need for lead extraction with a 2% complication rate and mortality of 2/1000 patients [5]. Thus, leads which can be easily removed and replaced are sorely need in the pediatric population. At the present time, small active fixation leads and ICD leads which have been treated to prevent tissue ingrowth tend to be more commonly used in the pediatric population because of these requirements.

2.1. Complex cardiac anatomy

Patients with congenital heart disease may have complex congenital anatomy which can complicate or even preclude transvenous devices. Patients may have obstructed venous pathways from prior central lines. They may have abnormal venous pathways, such as a left superior vena cava to coronary sinus (seen in up to 10% of patients with congenital heart disease) which may complicate or even preclude a transvenous system [6] (Fig. 3). Patients who have undergone an atrial baffling procedure (Senning or Mustard procedure) for transposition of the great arteries can develop stenosis of the SVC RA junction which can complicate pacemaker placement and require interventional stent placement prior to pacemaker implantation [7] (Fig. 4). The heart may not even be available via the transvenous system, such as in a patient with an extracardiac Fontan procedure, where the vena cavae bypass the heart entirely, and are attached directly to the pulmonary system. These patients require an epicardial system to access the atria.

Finally, patients with congenital heart disease often have intracardiac shunts. These patients have a greater than 2 fold increased risk of systemic thromboemboli with a transvenous system [8]. Disturbingly, aspirin or warfarin therapy was not protective against this risk. Thus transvenous leads should be avoided in the presence of an intracardiac shunt, and efforts to eliminate shunting should be pursued prior to transvenous lead implantation.

3. Lead position

The right ventricular apex is usually the most accessible position for lead placement in the ventricle both from the transvenous and the epicardial approach. Unfortunately, the electrical activation sequence produced when pacing in this location can have deleterious effect [9]. Chronic RV apical pacing causes RV dysynchronous contractions and decreases left ventricular function. This results in both acute and chronic deleterious hemodynamic effects, which can be especially problematic in patients who may require pacing for decades [10,11]. This has led several investigators to investigate alternative pacing sites which may more closely approximate a “normal” contraction sequence. His site pacing has been proposed as an alternative site as it allows for the most physiologic contraction sequence; however, technical issues, including lead stability, make this difficult to achieve in small children [12,13]. Recently, there has been a growing interest in left ventricular apical positioning of epicardial leads. Vanagt and colleagues have demonstrated that acute hemodynamic parameters are improved when compared to traditional RV epicardial pacing sites [13]. Studies are ongoing looking at chronic hemodynamic results with left ventricular pacing.

4. Venous obstruction

Venous obstruction following transvenous device placement is well described in the adult population with an incidence of 12% [14] (Fig. 5). In the pediatric population this incidence has been found to be anywhere between 13 and 20% [15,16]. There has been conflicting data as to the importance of patient size when compare to lead. This

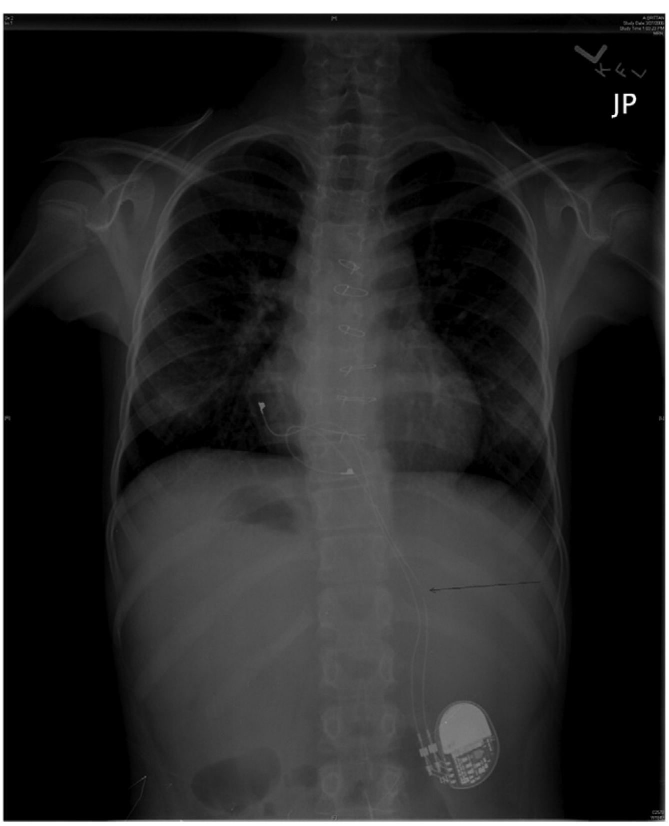

Fig. 2. Epicardial pacemaker lead break (arrow).

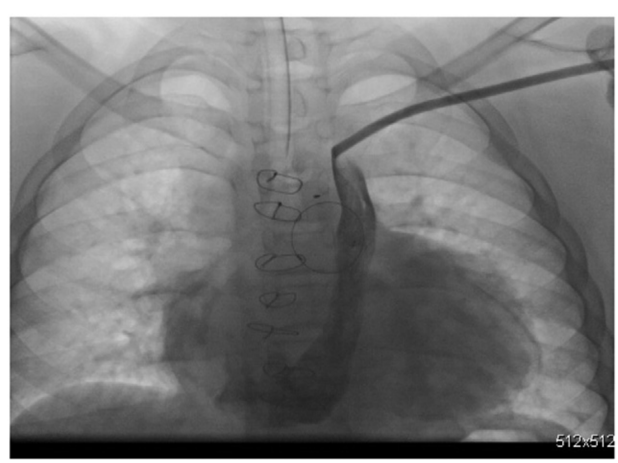

Fig. 3. Venogram of left superior vena cava draining to coronary sinus.