Since the advent of implantable cardioverter-defibrillators (ICDs) in the early 1980s, this therapy has been used in children. Indeed, one of the first subjects to receive a defibrillator in Mirowski’s landmark paper was a 16-year-old boy. This life-saving therapy quickly gained in popularity and its use has expanded to the point that over 5,000 ICDs are implanted per month in the United States. There have been multiple reports of the utility of this technology in the unique pediatric and congenital heart disease population. These studies have shown that indeed the ICD is as effective in treating a life-threatening arrhythmia in these patients as it is in the more typical patient with coronary artery disease.

However, as these devices have become more commonplace, we are becoming more aware of the “dark side” of defibrillator therapy. Inappropriate discharges, which can be problematic in the adult population, are more commonly seen in the pediatric and congenital heart disease populations, with an incidence of anywhere between 20% and 50% compared to the 17% seen in adults. In a direct comparison of pediatric versus adult ICDs placed in a single institution, with the same operators, there was a significantly higher incidence of complications, including infection and lead fractures.

As the pediatric electrophysiology community has become more comfortable with ICD therapy, practitioners have been willing to stretch the boundaries in order 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 (Figure 1). These novel techniques have allowed implantation of a defibrillator in children as young as a few months of age. However, these unique ICD configurations have an even higher chance of failing in the pediatric population, with a system survival rate of 50% at 3 years of follow-up.

The etiology of device failure in the pediatric and adult congenital population is multifactorial. High sinus rates, increased physical wear and tear, greater expected longevity, and, probably most importantly, growth have all been pointed to as reasons for lead failure and inappropriate discharges. These discharges can have serious psychosocial consequences, with patients describing post-traumatic stress syndrome and depression following an inappropriate discharge. Several studies of children have shown that children with ICDs often have higher levels of anxiety and worse quality of life scores than normal control subjects. Interestingly, no differences in quality of life were seen in patients who have had discharges versus those who have not.

There is also gathering evidence that inappropriate discharges can have serious physical consequences as well. Recent data from the SCD-HFT database has raised the level of concern regarding inappropriate defibrillator discharges, with a hazard ratio for death of 1.98 (1.2–3.0) with inappropriate shocks. Although it is important to acknowledge that the substrate in pediatric patients requiring ICD therapy is quite different from the SCD-HFT patient population, this finding does raise alarms, especially as (hopefully) our patients will have a much longer expected lifespan.

It is clear that ICDs in children require some major technical improvements. Suggestions have ranged from improved, stronger, smaller, non-thrombogenic leads that are easily removable to no leads at all; smaller
devices with longer battery life; better SVT/VT algorithms that have been tested in children; and less invasive epicardial delivery systems. However, there has been no real progress in any of these areas. At this date there is not a single pediatric-specific device available. Many reasons have been given by industry as well as by the regulatory community as to why this situation persists. These include a small patient population, technical difficulties in achieving our aims, financial considerations, and no mandate from the federal government. It is important that the pediatric community, device industry, and regulatory community work together to overcome these barriers. From easy fixes such as pediatric-specific tachycardia algorithms to the most complicated leadless device system, the pediatric community needs to be more closely involved in device development and testing both pre- and post marketing. It is time for change, and to truly innovate.

References


Figure 1: Chest radiograph of patient with a novel defibrillator configuration secondary to congenital heart disease. The implantable cardioverter-defibrillator coil is placed transcutaneously around the left ventricle.