Implantable Cardiacoverter Defibrillators in Children: Have We Reached the Threshold for Consensus?

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Editorial Comment

The loss of any family member is a devastating event; however, the sudden death of a child is particularly distressing, affecting not only the immediate family but also the whole community. As many such deaths are from ventricular arrhythmias, the advent of implantable cardioverter defibrillators (ICDs) represents a potentially life-saving therapy for those patients at greatest risk of sudden cardiac death (SCD). In this issue of the Journal, Alexander et al.1 describe a single-center’s experience with ICD therapy in 76 children and young adults with heart disease, the largest single-center “pediatric” ICD experience ever reported. There was a 28% incidence of appropriate, potentially life-saving ICD therapy, a 25% incidence of inappropriate discharge, and a 21% incidence of lead failure. The overall survival was 95%, with all 4 deaths occurring in patients with congenital heart disease (CHD).

Since the initial report by Mirowski et al.2 in 1980, the ICD has evolved from a large, cumbersome device able to deliver only high-energy shocks to a small, multiprogrammable device with dual-chamber (and biventricular) pacing capabilities. More than 60,000 devices are now implanted annually. As with most advances in medical technology, the application of ICDs in the pediatric population has been justified by extrapolating data from large trials in adult patients with diseases that are not prevalent in children. Although there have been numerous controlled, prospective trials demonstrating a clear advantage of ICD therapy over conventional therapy for prevention of SCD in adult populations (MADIT,3 AVID,4 MUSTT5), the indications for ICD use in pediatric patients with heart disease are neither well defined nor agreed upon. The 1998 ACC/AHA Guidelines for Implantation of Pacing Manufacturers and Antiarrhythmia Devices6 stated that the “indications for device therapy in pediatric patients are similar to those for arrhythmias in adults.” These recommendations were updated in 2002,7 although there remained no specific set of criteria for pediatric patients. A contributing factor to the lack of specific recommendations is the limited data regarding risk stratification in pediatrics.8,9 Although risk stratification data have been proposed for certain diagnoses prone to SCD (i.e., long QT syndrome, Brugada syndrome, hypertrophic cardiomyopathy), these data do not necessarily pertain to pediatric patients and are based on experiences at tertiary care centers.10-12

Although there is general consensus that young survivors of aborted SCD would benefit from ICD therapy, the reported experience in pediatric patients is limited to retrospective reviews.13-17 A major hurdle for arriving at such a consensus is the relatively limited number of pediatric patients undergoing ICD placement, with <1% of annual implants in those younger than 21 years. As appropriately pointed out by Alexander et al.1 it would require >950 ICD patients (with equal number of controls) to demonstrate a 5-year survival improvement from 85% to 90%. However, the low rate of appropriate (28%) and near-equal rate of inappropriate (25%) discharges in this series highlights the urgent need for improved selection criteria.

What is the acceptable and optimal rate of appropriate ICD discharge? A rate of 100% means that patients who potentially might benefit from this therapy are omitted to avoid any uncertainty. Would implanting devices in those with minimal risk of SCD be unreasonable if specificity was so high that no inappropriate discharges occurred? These questions are central to the expanded utilization of these devices in young patients and must await further descriptive data, as a placebo-controlled trial would be unethical.

As the indications for ICD therapy in pediatrics have broadened, its use has become more widespread, with devices being implanted in children with primary electrical disorders or cardiomyopathy, and in certain subsets of CHD known to have an increased incidence of SCD. When evaluating the current rates of appropriate ICD discharge, one must take into account the dramatic change that has occurred in patient selection over the past decade. In 1993, Silka et al.13 reported a 59% incidence of appropriate discharges in a multi-institutional early report of 125 pediatric patients. However, the majority of the patients were selected as survivors of SCD (76%), and very few (18%) were patients with CHD. In the current report, on the other hand, only 32% had a prior history of cardiac arrest, and 42% had a history of CHD. Those patients without a history or cardiac arrest were stratified with a combination of family history of SCD, patient symptoms, clinical findings, and response to programmed electrical stimulation.1 Those with primary electrical disease had the highest incidence of appropriate shocks (44%) and cardiac arrest at presentation (48%).3 The challenge in the heterogeneous pediatric population will be to determine which patients, aside from those with a history of aborted SCD, will benefit from ICD therapy.

Other considerations in pediatric patients include the psychosocial impact, small patient size, and higher incidence of system failure compared to adult patients. System malfunctions invariably lead to further invasive procedures, may result in inappropriate discharge, and/or potentially leave a child unprotected. A higher incidence of pediatric infection and lead malfunction was reported in an earlier single-center comparison of pediatric and adult patients.18 Small patient
size contributed significantly to the complication rate in the current series as well. These findings also highlight the need for continued improvement in device technology. Although further miniaturization has allowed for device implantation in smaller patients, manufacturers share a responsibility to further improve lead delivery, durability, and longevity. Additionally, there is a need for continued refinements in tachycardia detection and discrimination because inappropriate discharges may have a devastating effect on these young and psychologically vulnerable patients.

Pediatric electrophysiologists must develop a consensus on indications for implantation of ICDs and agree upon criteria for monitoring the impact of this therapy. A comprehensive registry of young ICD patients has been created and currently is in its early stages through the Pediatric Electrophysiology Society. A similar registry was developed in 1991 to evaluate the outcomes of radiofrequency catheter ablation in young patients. By 1999, >6,000 pediatric patients had been enrolled into the Pediatric Radiofrequency Ablation Registry. This registry not only helped define the indications and success/complication rates of this procedure but also contributed to the development of catheter advances and diagnostic strategies. The stakes in SCD are much greater while the patient volumes are lower, emphasizing the need for cooperative multicenter studies among pediatric cardiologists to define the indications and expected outcomes of ICD therapy in young patients with heart disease.

Despite the shortcomings of ICD therapy in young patients, this technology has had a measurable benefit in the survival of pediatric patients at risk for SCD. Incorporating important studies such as those of Alexander et al. with a comprehensive prospective registry, it should be possible to further delineate the indications for ICD therapy and improve survival in these high-risk patients.

References