Pacemakers and Other Cardiac Implantable Devices in Children
Learning From Practice Variation Including Outliers in the Electronic Age

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... Two roads diverged in a wood, and I—
I took the one less traveled by,
And that has made all the difference.


The first myocardial pacing wire was implanted ≈6 decades ago in 1957 by cardiothoracic surgeon Dr C. Walton Lillehei.1 The recipient of this epicardial therapeutic cardiac electronic device was a 3-year-old girl with complete heart block after repair of tetralogy of Fallot. Since then, the science and practice of cardiac implantable electronic device (CIED) use for diagnosis and treatment of heart rhythm and heart failure disorders in children and adults has grown exponentially. Indeed, in 2016, the tables have turned, whereas Dr Lillehei and other pioneers in cardiology, cardiothoracic surgery, and CIED development faced a void of options to diagnose and treat their patients with life-threatening rhythm problems, modern clinicians, in a new era characterized by “should we” rather than “can we,” face the relatively innocuous dilemma of what type of CIED to choose.

See Article by Konta et al

Scientific publications like the study by Konta et al2 in this issue of *Circulation: Arrhythmia and Electrophysiology* add valuable information to the body of knowledge that guides our choice of CIED for individual patients. Despite >5 decades of use in children and congenital heart disease, the decision process is not straightforward. Clinical reports describing outcomes of CIED including pacemakers for children and adult patients with congenital heart disease often provide conflicting results. Prospective randomized trials or registries of CIED use in children and congenital heart disease are rare, and nearly all published data come from retrospective studies like the current report from Konta et al.2 As a result, guidelines that address appropriate indications for pacemaker and implantable cardioverter defibrillator use in children and congenital heart disease are supported by suboptimal levels of evidence (only B and C in the most recent American College of Cardiology (ACC)/American Heart Association/The Heart Rhythm Society 2008 Guidelines).3

Another consequence of suboptimal evidence in the area of pacemaker use for children and congenital heart disease is a lack of published guidelines beyond basic pacing indications. The 2008 American Heart Association/ACC/The Heart Rhythm Society writing committee states “additional details that need to be considered in pacemaker implantation in young patients include risk of paradoxical embolism due to thrombus formation on an endocardial lead system in the presence of residual intracardiac defects and the lifelong need for permanent cardiac pacing. Decisions about pacemaker implantation must also take into account the implantation technique (transvenous versus epicardial), with preservation of vascular access at a young age a primary objective,” but the committee stops short of detailed guidelines addressing these aspects of practice.1 Konta et al2 recognize that their practice is unusual and that the high rate of venous occlusion in small children after endocardial lead placement has lead a majority of pediatric electrophysiologists to favor epicardial pacing and defibrillation leads for patients weighing <10 to 15 kg.4,5 That said, the outcomes of endocardial pacing system placement reported by Konta et al2 are unusually favorable for patients weighing >5 to <10 kg when compared with prior studies, documenting a low 8% incidence of subclavian vein occlusion in 13 patients at a median of 9.2 years (range, 3 months to 12.9 years). The incidence of subclavian vein occlusion reported by Konta et al2 for patients weighing <5 kg is closer to prior publications at 70% in 13 patients at a median of 9.2 years (range, 3 months to 12.9 years).

In addition to a lower rate of venous occlusion compared with previous reports, Konta et al2 document favorable longevity of endocardial pacing leads in small children with 100% lead survival in 22 patients at 3 years, 95% at 5 years, and 73% at 8 years. This rate of lead survival is better than the previously reported rates of epicardial pacing lead survival for children of all ages: 58% to 83% at 5 years.9,10 Finally, despite the fact that they are treating children <10 kg, Konta et al2 report a rate of procedural and device complications similar to many other series of endocardial pacemaker implantation in larger children including acute lead dislodgment and infection. This may be, in part, because of the skill of 1 implanter who performed all endocardial pacemaker implants in the series since 1990—24 of the 27 years reported. It is also possible that fine details of the implant technique performed by this solo implanter, including methods for venous access, lead tie down, vascular maneuvers, lead slack placement,
have lead to the superior performance for endocardial leads in the very young patients in this series. Konta et al do not affirm or refute cases of thrombosis or thromboembolism beyond the subclavian vein, including no report of stroke or systemic thromboembolism, despite the known risk for events with endocardial leads in the reported patient population that includes congenital heart disease and patent foramen ovale (patent foramen ovale was not addressed in the report by Konta et al, but presumably present in most newborns and at least 25% of older children with normal anatomy) with a potential for endocardial right to left shunt.11

Overall, Konta et al are to be congratulated; they have taken the road less travelled and their smallest patients have benefitted from their expertise and fortitude with excellent outcomes of long-term endocardial cardiac pacing. If this is truly the case, and there is potential for superior outcomes of endocardial cardiac pacemaker in the smallest children, especially those weighing >5 kg, why is their road less travelled? The answer lies in the quality of shared knowledge in the area of pediatric and congenital electrophysiology. We treat rare cardiac diseases with a limited number of patients at any one pediatric and congenital heart center. Even the largest congenital heart centers do not have enough volume of patients or enough practice variation to determine optimal CIED use for each type of pediatric or congenital heart patient.

Many fields of medicine, including cardiology and cardiothoracic surgery, have turned to prospective collaborative clinical registries to understand fine details on optimal care for patients with rare and orphan diseases such as pediatric and congenital heart disease. A prime example of such collaboration is the Data Center of the Congenital Heart Surgeons’ Society, a group of pediatric heart surgeons now representing 65 institutions that treat patients with congenital heart defects. The history of the group goes back to the early 1970s, when congenital cardiothoracic surgeons started to meet annually to relate their early experience operating on children with congenital heart defects. Founding members Dr Kirklin and Dr Blackstone recognized that the occurrence of congenital heart disease is so low that only by combining the experience of the Congenital Heart Surgeons’ Society members through the Data Center could they improve their ability to determine the best methods for treating congenital patients. Since the first prospective study on operative treatment for D-transposition of the great arteries started enrolling in 1985, the Congenital Heart Surgeons’ Society has been extremely productive having published dozens of seminal reports about surgical repair of congenital heart disease.

Recently, the Congenital Heart Surgeons’ Society began public reporting for participating centers, an important step in the process of quality improvement and transparency in the diagnosis and treatment of patients with congenital heart disease.12 A similar effort in the area of pediatric and congenital interventional cardiology, aimed primarily at quality improvement, was launched in 2011 through the efforts of the Congenital Heart Disease Committee of the Society of Cardiovascular Angiography and Interventions and the ACC National Cardiac Data Registry (NCDR). The Web-based electronic Improving Pediatric and Adult Congenital Treatment (IMPACT) Registry started to enroll children and adult patients with congenital heart disease in 2011 and now includes data from 91 pediatric and congenital heart centers. Publications from the IMPACT registry have been forthcoming and shed important new light on treatment outcomes for interventional cardiac catheter–based procedures for children and patients with congenital heart disease.13,14

ACES (the Pediatric and Congenital Electrophysiology Society) has supported effective international collaboration between pediatric and congenital electrophysiologists since the early 1980s (members include authors of the report by Konta et al in this issue of Circulation: Arrhythmia and Electrophysiology). Many leaders in the field of pediatric and congenital electrophysiology have sponsored effective voluntary registries to answer basic questions about optimal treatment for our patients including cardiac catheter ablation and CIED therapies.15–17 In collaboration with Pediatric and Congenital Electrophysiology Society, the ACC NCDR has supported the addition of a pediatric and adult congenital ablation module to the electronic IMPACT Registry, because of go live in late 2016. After the ablation module, Pediatric and Congenital Electrophysiology Society and ACC NCDR leadership have made preliminary plans for a comprehensive pediatric and adult congenital CIED module to be added to a later version of the IMPACT registry. Indeed, there have been some data generated by the current NCDR implantable cardioverter defibrillator Registry for pediatric and congenital patients, but the utility has been limited because of a low participation rate, (primarily because of the cost of supporting a second NCDR registry at pediatric and congenital heart centers), a limited number of data fields related to pediatrics and congenital heart disease, and no data from CIEDs other than implantable cardioverter defibrillators.18–20 The new IMPACT registry module will add yet other electronic dimension beyond the current practice for CIED use for children, allowing us to take advantage of current practice variation and learn from outliers who have taken the road less travelled in the field of pediatric and congenital CIED therapy. The ultimate goal of our efforts is to provide improved and optimal outcomes for our pediatric and adult congenital patients who required CIED therapy for diagnosis and treatment of a rare disease; our patients who have no choice but to take the road less travelled.

Disclosures
None.

References


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