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STATE-OF-THE-ART REVIEW

The Current State and Future Potential of Pediatric and Congenital Electrophysiology

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ABSTRACT

Pediatric electrophysiologists specialize in the diagnosis and treatment of rhythm abnormalities in pediatric, congenital heart disease, and inherited arrhythmia syndrome patients. The field originated out of the unique knowledge base that rhythm management in young patients required. In the 1970s, pediatric electrophysiology was recognized as a distinct cardiac subspecialty and it has evolved rapidly since that time. Despite the considerable growth in personnel, technology, and complexity that the field has undergone, further opportunities to progress pediatric electrophysiology exist. In this review, we highlight some of the clinical focus of pediatric and adult congenital electrophysiologists to date and identify areas within this specialty where the pediatric and congenital electrophysiology community could come together in order to drive improvements in rhythm management for patients. (J Am Coll Cardiol EP 2017;3:195–206)

The field of pediatric electrophysiology (EP) developed in the latter half of the 20th century and has evolved into a fully recognized subspecialty within pediatric cardiology. Pediatric EP has an intimate relationship with the field of adult EP, built on the foundation of a common physiology and fostered by shared clinical interests. Familial and genetic arrhythmia syndromes and adult congenital heart disease, in which sudden death and arrhythmia are important clinical challenges, represent 2 expanding intersects of adult and pediatric care models. Although there are differences in the type and prevalence of arrhythmia substrates on which each field focuses, the underlying physiology and pathophysiology, mechanisms, and principles are similar. Similarly, the technologies being developed for cardiac rhythm devices and ablation are shared.

Although the mainstay of adult EP is prevalent diseases with older age at onset (e.g., atrial fibrillation, ischemic heart disease, heart failure, and their sequelae), pediatric EP encompasses a broad collection of individually uncommon and rare disorders. These unique disorders contribute to some of the

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ABBREVIATIONS AND ACRONYMS

ACHD = adult congenital heart disease

AVN = atrioventricular node

CCTGA = corrected transposition of the great arteries

CHD = congenital heart disease

CRT = cardiac resynchronization therapy

EP = electrophysiology

HF = heart failure

ICD = implantable cardioverter-defibrillator

IHRD = inherited heart rhythm disorders

JET = junctional ectopic tachycardia

LV = left ventricle

MAP-IT = Multi-Centre Pediatric and Adult Congenital EP Quality

RFCA = radiofrequency catheter ablation

RV = right ventricle

SIDS = Sudden infant death syndrome

SVT = supraventricular tachycardia

TOF = tetralogy of Fallot

challenges of pediatric EP, including the lack of high-quality evidence supported by large clinical trials, the issue of orphan drugs, and the need to adopt primarily adult-based technology.

Typically, pediatric electrophysiologists dedicate an additional 1 to 2 years of training after a pediatric cardiology fellowship. They work predominantly in academic centers and constitute the majority of members in the Pediatric and Congenital Electrophysiology Society (PACES). PACES began in 1980 (1) as an informal collaborative and has enjoyed continuous growth over the past 35 years. PACES has been prominent in pediatric and congenital EP in developing clinical standards and conducting collaborative research. Internet-based international collaborations have become a common research methodology, and PACES and its members have produced several multicenter, multinational publications in the past decade (2). Against this background of discovery and collaboration, there is an opportunity and an obligation to come together to lead developments in the field of pediatric and congenital EP. In this commentary, we highlight the current activities of pediatric and adult congenital electrophysiologists and identify areas that could benefit from intensified efforts by this community in order to drive improvements in clinical care and to advance our field (**Central Illustration**).

ATRIOVENTRICULAR NODE

The atrioventricular node (AVN) was first described over 100 years ago, and its complex physiology remains incompletely understood. Anatomic variability and autonomic factors contribute to the challenges in elucidating the physiology of the AVN in health and disease. Added complexity comes from the dual electrophysiological character of the AVN as it is both a conductor of impulses and an oscillator.

POST-OPERATIVE AVN COMPLICATIONS. Following heart surgery close or adjacent to the AVN, the transient but malignant arrhythmia junctional ectopic tachycardia (JET) may arise secondary to enhanced automaticity. Genetic determinants of JET have been reported based on rare cases of congenital and familial JET (3,4). JET is not commonly observed in adult patients, likely owing to the maturational changes within the AVN that have been demonstrated in animal studies (5). These changes include a reduced capacity for rapid antegrade conduction and development of multiple electrical pathways (5). The latter can support a re-entrant arrhythmia, atrioventricular nodal re-entrant tachycardia. Atrioventricular nodal re-entrant tachycardia is uncommon in infants, although it increases in adolescence, and is a common indication for ablation therapy in adults (6). To date, heart block remains one of the most frequent complications of open heart surgery for congenital heart disease (CHD) (7,8). In rare cases, heart block may have a congenital origin, most commonly secondary to maternal antibody exposure, and will often require pacemaker insertion in neonates and infants.

FUTURE CONSIDERATIONS. Although pediatric electrophysiologists expend considerable time and resources treating conditions associated with AVN pathology, relatively little research has been undertaken to improve our understanding of the physiology and pathophysiology of the developing AVN. Recent evidence in animal models suggests ischemia-reperfusion time plays an important role in the pathogenesis of post-operative arrhythmias, including JET (9). However, we remain limited in our ability to predict and prevent post-operative JET (10,11) and are unable to decrease the incidence of post-operative heart block. It is important that we switch our focus away from accepting these post-operative complications as inevitable consequences of cardiac operations and, instead, work to prevent their occurrence by elucidating their pathophysiology through further partnerships with basic scientists and researchers; this will aid in continued development of AVN models and potentially help identify markers for AVN cells.

CARDIAC RHYTHM DEVICES

Pediatric EP has its roots in cardiac pacing and post-operative heart block (2). Although most post-operative heart block cases are transient, 1% to 2% require permanent pacing (12). Rates of lead failure in pediatric patients are high, and current pacemaker technology is not well adapted for small or growing individuals (13). Although the present technology has become an acceptable norm, a significant obstacle continues to be access to the heart and need for multiple invasive procedures. Leadless pacemakers have shown early success in adults, with similar complication rates and improved battery longevity compared to conventional pacemakers (14); however, there has been no direct comparison between the 2, and there is insufficient data for long-term efficacy