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STATE OF THE ART REVIEW

Advancing the science of management of arrhythmic disease in children and adult congenital heart disease patients within the last 25 years

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Abstract

This review article reflects how publications in EP Europace have contributed to advancing the science of management of arrhythmic disease in children and adult patients with congenital heart disease within the last 25 years. A special focus is directed to congenital atrioventricular (AV) block, the use of pacemakers, cardiac resynchronization therapy devices, and implantable cardioverter defibrillators in the young with and without congenital heart disease, Wolff–Parkinson–White syndrome, mapping and ablation technology, and understanding of cardiac genomics to untangle arrhythmic sudden death in the young.

Keywords

Paediatric electrophysiology • Adult congenital heart disease • Congenital AV block • Pacemaker • ICD • WPW syndrome • Sudden cardiac death • Mapping and ablation • Supraventricular and ventricular tachycardia

What's new?

This review article reflects how publications in EP Europace have contributed to advancing the science of management of arrhythmic disease in children and adult patients with congenital heart disease within the last 25 years. Special focus is directed to congenital AV block, the use of pacemakers, cardiac resynchronization therapy devices, and implantable cardioverter defibrillators in the young with and without congenital heart disease, Wolff–Parkinson–White syndrome, mapping and ablation technology, and understanding of cardiac genomics to untangle arrhythmic sudden death in the young.

Introduction

The field of paediatric and adult congenital electrophysiology encompasses a large patient age group and an enormous disease spectrum. Over the 25 years since it started publication, Europace has helped to navigate this complexity. In this time, the field has expanded along with an increasing number of adult survivors with congenital heart disease (CHD), many of whom have associated tachyarrhythmias, sinus node, or AV node disease. There has been a significant expansion in device technology and the application of pacemakers and implantable cardioverter defibrillators (ICDs) to expanding populations. Mapping and ablation technology has allowed for the cure of a growing number of arrhythmias with increased success and safety. Finally, our expanding understanding of cardiac genomics has helped us untangle arrhythmic sudden death in the young. In this issue, we will explore the contribution of Europace to advancing the science of the management of arrhythmic disease in children and adult CHD (ACHD) patients.

Congenital AV block

Possibly the most difficult population that paediatric electrophysiologists deal with are newborns with congenital complete AV block (CCAVB). Although rare, 1:14 000–20 000 live births,[1,2] this is the most common indication for pacemaker implantation in the very young. Once embarked upon, pacing in this population means a lifetime of device therapy and the myriad of complications and consequences arising out of this.

The optimal timing of pacemaker implantation in the CCAVB population remains an issue of debate. In the absence of symptoms, avoiding pacing as long as it is safe makes sense. However, natural history studies have demonstrated progressive left ventricular dysfunction and mitral insufficiency with cardiovascular mortality in the fourth or fifth decade in patients with CCAVB without a pacemaker.[3] Contemporary studies on patients with isolated CCAVB who do not undergo pacing remain a knowledge gap. In a 2007 Europace publication, Vukomanovic *et al.*[4]

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looked at the Holter-determined mean heart rate in predicting symptoms of heart failure and Adams–Stokes attacks (syncope) in children with CCAVB. They found that the maximum heart rate was a better predictor of symptom development than mean daytime heart rate as were pauses of at least twice the basic cycle length. Although heart failure and syncope can develop at any age, those <8 years had an increased risk for heart failure and older children were more likely to have syncope. The authors advocated for 24 h Holter once or twice a year for risk stratification and decision-making for a pacemaker implantation. Although the average ventricular rate in newborns and infants with CCAVB provides an objective measure regarding the decision for pacemaker implantation, additional factors may equally influence the decision/timing of pacemaker implant.[5] These include size, CHD, ventricular function, and other comorbidities.

Pacemaker and resynchronization devices

Safety and efficacy of long-term/life-long cardiac pacing in infants and children for complete AV block and sinus node dysfunction have been a matter of debate since the 1980s. Despite the fact that endocardial pacing in the young is the more feasible option,[6] epicardial pacing is still required in patients with concomitant cardiac surgery, intracardiac shunting, difficult or absent venous access, and small patient size.[7,8]

Publications in EP Europace contributed significantly towards improving knowledge and quality of care in this field. A single-centre study of 287 patients with CHD requiring cardiac pacing showed a significantly higher risk of failure of epicardial leads when compared with endocardial systems during a median follow-up of 5 years. Younger age at implantation was a risk factor for complications at follow-up.[9] To improve results in epicardial pacing, steroid-eluting epicardial pacemaker implantation in young children was shown to be associated with a satisfactory outcome, while major complications were related to lead fracture.[10] AutoCapture®-controlled epicardial pacing and Ventricular Capture Management™ were safe and effective and able to prolong calculated battery life up to 15%.[11–13] Despite the disadvantages of epicardial pacing, data from the Czech Republic published in Europace in 2012 in patients <18 years of age undergoing permanent epicardial pacemaker implantation showed the probability of continued epicardial pacing at 93 and 76% after 5 and 10 years, respectively. The use of bipolar steroid-eluting electrodes and the AutoCapture® feature decreased the risk of surgical reintervention and battery depletion and allowed transvenous pacing to be deferred to an older age.[14]

Choosing the optimal pacing site for chronic ventricular pacing has recently been identified to be of major impact in preserving ventricular function.[15,16] The detrimental effects of right ventricular (RV) apical pacing in the young resulting in pacing-induced cardiomyopathy have well been described.[17,18] The results of single-site left ventricular epicardial pacing published in Europace demonstrated preserved left ventricular synchrony and function in paediatric patients when compared with RV pacing[19,20] and is now accepted as the preferred site for epicardial pacing.[8]

The implantation of a permanent pacemaker in children and adolescents usually has a favourable outcome, but children might be more prone to complications because of their active lifestyle, higher frequency of traumatic events, and infections that affect the pacing system. Pacing leads remain the 'weakest link' of the pacing system and long-term complications are mainly lead-related.[21] In a series of 292 children spanning 20 years of pacing, almost 20 years ago, pacemaker-related deaths were not seen, but serious complications needing re-operations or a revision of the system occurred. Early complications were related to infections (2%) and, in endocardial pacing, to haemothorax or vascular haemorrhage (3.5%) or to lead dislodgement (5%).

In 2010, Silvetti *et al.*[22] analysed risk factors for the development of dilated cardiomyopathy in the paced infant and alerted us to the fact that high-rate ventricular pacing potentiates the development of dyssynchrony and subsequent dilated cardiomyopathy. Those with DDD pacing were more likely to develop cardiomyopathy than those with VVI pacing, and cardiomyopathy did not occur in infants in whom the ventricular pacing rate was slow and gradually increased before switching to DDD pacing. Sometimes less is in fact better.

Cardiac resynchronization therapy

Cardiac resynchronization therapy (CRT) has evolved as an important adjunct to medical therapy in adult patients with heart failure and reduced ejection fraction (EF), allowing robust recommendations.[23,24] Data in the young have shown that patients with RV-pacing-induced cardiomyopathy are the best responders for CRT as well as patients with a failing systemic left ventricle and left bundle branch block pattern.[25–27] Up to now, research on CRT in young adults with CHD has not reached conclusive results, precluding evidence-based recommendations.[28,29] This particularly applies to patients with repaired tetralogy of Fallot (TOF), a systemic RV and those with a single ventricle.[30,31] This knowledge gap is due to the limited number of patients with different underlying substrates. Data published in Europace on CRT in patients with ACHD were limited to 48 patients and 11 individuals, resulting in a success rate comparable with those with acquired heart disease.[32,33] Another report on the effects of CRT on echocardiographic indices, functional capacity, and clinical outcome in systemic RV patients was based on a total of only seven patients.[34] Cardiac resynchronization therapy for the failing systemic RV is a complex issue when compared with left ventricular CRT. This is due to different RV structure, structural tricuspid valve regurgitation vs. functional mitral valve regurgitation in systemic left ventricle, and decreased myocardial perfusion reserve. It is of note that promising results were published on CRT in young adults with congenitally corrected transposition of the great arteries (cc-TGA),[34,35] where access to the coronary sinus can allow for traditional lead placement.

Right heart failure combined with the right bundle branch block pattern is an important cause of late morbidity and mortality in patients with repaired TOF. This topic gained considerable attention in Europace. First attempts at applying atrio-synchronized RV pacing failed to result in significant haemodynamic improvement.[36] Detailed electroanatomical mapping of RV activation in postoperative patients with TOF helped to improve haemodynamics by targeting single- and dual-site RV pacings,[37,38] but data remain limited.

Conduction system pacing in congenital heart disease

Traditional CRT via a lead implanted into the coronary sinus may not be feasible in patients with CHD due to associated lesions or prior surgery. Conduction system pacing has been shown to be equivalent to traditional CRT in patients with CHD.[39] Successful left bundle branch area pacing in this patient population focusing on techniques for implantation has recently been reported in EP Europace.[40]

Atrial pacing in preventing atrial arrhythmias in patients with congenital heart disease

Indications for pacemaker implantation have been updated recently by the European Society of Cardiology.[24] Increasing the heart rate by

permanent atrial pacing to prevent atrial arrythmias may be considered,[41] but as published in Europace in 2013, in contrast to the general population, atrial-based pacing was not associated with a lower incidence of arrhythmias in patients with ACHD.[42]

Wolff–Parkinson–White syndrome

Wolff–Parkinson–White (WPW) syndrome is considered by many electrophysiologists, the classical arrhythmic disease. A famous metaphor by James calls the WPW syndrome 'the Rosetta stone of electrocardiography, since a full understanding of all its features and their possible mechanisms encompasses many fundamentally important principles'.[43] Wolff–Parkinson–White has fostered the correct interpretation of circus movement as the cause of tachycardia and can therefore indeed be compared with the deciphering of hieroglyphic writing by Champollion in 1822 with the aid of the Rosetta stone. In a 2009 Europace editorial, Lüderitz[44] described this symptom complex as characterized by a double excitation of the heart chambers caused by pre-excitation along an accessory excitation pathway bypassing the AV node. The additional connection fulfils the anatomic and functional requirements for circus movement. Clinically, this usually takes the form of supraventricular tachycardia (SVT) but an individual with WPW can have a variety of manifestations from asymptomatic pre-excitation to sudden death. Although the most common symptom is SVT, some patients with WPW are at risk for sudden death. Although this risk is small, it is front loaded on the young.[45]

In 25 years, since the inaugural issue of Europace, there has been a change in the management of WPW. Much has happened to make ablation safer and more successful. In the 2009 European Heart Rhythm Association (EHRA) survey on ventricular pre-excitation, 69% of centres would have referred patients with asymptomatic pre-excitation for risk stratification[46] and in a subsequent similar EHRA survey, a young asymptomatic patient would be risk stratified or ablated in 84% of the 58 centres covering 20 countries.[47] All were high-volume ablation centres and, in this group, a younger person with an asymptomatic WPW pattern had a higher likelihood of being risk stratified or receiving ablation therapy compared with an older subject. Paediatric electrophysiologists agree that symptomatic WPW requires catheter ablation once size limits have been met.[48] There remains debate about the management of an asymptomatic patient with pre-excitation. Paediatric and adult electrophysiologists differ in this with paediatric electrophysiologists increasingly advocating ablation in children without symptoms.[49] This is in part due to concerns that a life-threatening arrhythmia may be the first symptom in children with WPW.[50]

Predicting risk in asymptomatic children with pre-excitation is difficult. Using transoesophageal electrophysiology studies, Di Mambro *et al.*[51] assessed the accessory pathway effective refractory periods (APERPs) and tachycardia inducibility in 124 children aged 4–18 years. Over half were asymptomatic and were compared with the group with symptoms. At baseline, there was no difference in APERP, atrioventricular reentry tachycardia, or atrial fibrillation (AF) inducibility between those with and without symptoms. Isoproterenol or exercise testing made the inducibility of SVT more likely in those with symptoms. However, conduction properties of accessory pathways were not different between the groups during isoproterenol or exercise testing.

A successful catheter ablation cures WPW but are there any long-term consequences? The traditional belief is that ablation prevents recurrences of AF. Borregaard *et al.*[52] studied 362 patients aged 0–80 years who had undergone an ablation for WPW with 16% aged 18 years or younger. The risk of AF after ablation remained higher in the WPW group than in a background population, whereas the long-term mortality was not different. It is still unclear why the risk of AF remains after ablation of WPW. The patients with WPW did have more comorbidity with a higher prevalence of heart failure, hypertension, valvular heart disease, ischaemic heart disease, and CHD, all of which are associated with AF. However, the risk of AF remained higher even after adjustment for these comorbidities. Atrial fibrillation before ablation was an independent risk factor for AF after ablation. This suggests that electrophysiological abnormalities present before ablation at least partially determine the occurrence of AF afterward. Atrial fibrillation in WPW before the ablation might lead to atrial remodelling, which begets further AF after the ablation. There was an age-dependent risk of post-ablation AF in the WPW group. Patients with WPW aged 50 years or more had a significantly higher risk of post-ablation AF than the younger group (hazard ratio: 9.79 and confidence interval: 4.29–22.36). Is there an argument here for earlier catheter ablation as a means to decrease the lifetime burden of AF in this population?

In the 1970s, it was first noted that WPW can also result in changes in interventricular septal motion, as noted on echocardiography.[53] This was underappreciated until catheter ablation offered a cure for WPW. In a 2007 Europace article, Tomaske *et al.*[54] focused our attention on this negative consequence of WPW. They assessed the effect of ablation of the right septal and posteroseptal accessory pathways on left ventricular function. This pivotal article looked at 34 patients who, in the absence of incessant SVT, had reversible changes in left ventricular function after ablation. Prior to ablation of a right septal or posteroseptal accessory pathway, 12% of the patients had clinical heart failure, while mean left ventricular EF was moderately impaired. After ablation, a rapid and significant improvement in left ventricular function was noted. Therefore, in addition to the potential risk of a life-threatening arrhythmia, the resolution or prevention of left ventricular dysfunction is yet another reason to eliminate WPW.

To what extent and when does the pre-excitation-driven dyssynchronous contraction occur in the setting of a child or young person with WPW? Dai *et al.*[55] examined the influence of the accessory pathway and pathway location on ventricular wall motion and left ventricular function. They found that right septal and right free-wall accessory pathways were associated with left ventricular dysfunction and left ventricular dilation. This was not seen in patients with left-sided pathways or in controls. Left ventricular dyssynchrony and abnormal interventricular septal motion were thought to be the genesis of the dysfunction and chamber enlargement.

In a compelling 2022 Europace article, Ksiazczyk *et al.*[56] identified that physical performance as measured by cardiopulmonary exercise testing (CPET) was worse in children with WPW even in the absence of symptoms or tachycardia. They found that key CPET parameters of physical performance were affected by pre-excitation. Cardiopulmonary exercise testing data showed significantly lower VO_2 peak and O_2 pulse in patients with WPW compared with healthy controls. Patients with persistent pre-excitation had lower VO_2 peak, anaerobic threshold, and O_2 pulse when compared with those with loss of pre-excitation with exercise.

Adult congenital heart disease

The population of patients with CHD is increasing, and more patients reach adulthood with more complex disease. A significant portion of these patients will suffer from arrhythmias due to the underlying CHD or as a sequela of interventional or surgical treatment. The medical community will encounter an increasing challenge, and to address these, in 2018, Europace published a consensus paper concerning the arrhythmia burden in the ACHD population.[28]

Arrhythmias are a major cause of morbidity and mortality in patients with ACHD. Patients with ACHD can be as complex as the palliated single ventricle patient or as seemingly simple as a repaired atrial septal defect (ASD). In 2003, Mantovan *et al.*[57] reported on 136 consecutive young adults who had undergone ASD repair. Sustained supraventricular arrhythmias occurred in 12 patients before surgery and 11 had AF.

The occurrence of arrhythmias significantly correlated with the presence of coexisting heart disease, most of which was mitral valve disease and older age at surgery. After surgery, sustained arrhythmias were recorded in 16 patients, 11 of which were AF, 4 intraatrial reentrant tachycardia (IART), and 1 atrioventricular nodal reentrant tachycardia (AVNRT). There was a correlation between pre- and postoperative arrhythmia. This was an early identification that AF is the most frequent form of arrhythmia before and after surgical closure of ASD in patients with ACHD and relates to age at the time of repair and coexisting heart disease.

Although IART is the most frequent arrhythmia in the younger ACHD population, the incidence of AF is increasing with patients' age, surpassing IART in people >50 years.[58] Unique anatomy and physiology, a high diversity of corrective surgeries and associated comorbidities add complexity to the treatment of this challenging arrhythmia.

Paroxysmal AF is known to occur in patients with unclosed ASD. In a 2019 issue of Europace, Nakagawa *et al.*[59] sought to assess the efficacy of catheter ablation compared with transcatheter ASD closure alone for treating pre-existing paroxysmal AF in patients with ASD. Among 908 patients who underwent transcatheter ASD closure, 50 patients (mean age 63 ± 12 years) with paroxysmal AF were assessed. The AF-free survival rate was significantly higher for patients with an ablation compared with those with ASD closure alone. Catheter ablation prior to ASD closure substantially suppressed AF recurrence over the long term. A combination of ablation and transcatheter ASD closure seems to be a feasible treatment strategy for paroxysmal AF in patients with ASD.

Recurrent atrial tachycardia is common after repair of many types of CHD, and surgical ablation with a maze procedure represents a potential treatment strategy. A 2019 Europace study reported on a single-centre's 19 years' experience with maze surgery in patients with CHD.[60] In this study, the maze procedure was classified as therapeutic if the patient demonstrated preoperative atrial arrhythmias, or as prophylactic if done because the patient was considered high risk for postoperative arrhythmias. A maze was performed on 137 subjects in the therapeutic group and 29 in the prophylactic group. Although IART was the most common arrhythmia, 18% of patients had AF and 20% had both. The most common CHD lesion was a single ventricle in the therapeutic group (27%) and Ebstein's anomaly of the tricuspid valve in the prophylactic group. The procedures consisted of a right atrial maze in 63%, a left atrial maze in 4%, and a bilateral maze in 33%. There were no direct complications or mortality related to the maze procedure. For the therapeutic group, freedom from arrhythmias was 82 and 67% at 1 and 5 years, respectively. A younger age at surgery was correlated with a lower long-term recurrence risk. Maze procedure at the time of an elective anatomic surgery was reasonably effective to prevent and treat atrial arrhythmias in patients with CHD in the short- and mid-term, with low morbidity and mortality.

Ventricular arrhythmias and implantable cardioverter defibrillators in the young and in patients with congenital heart disease

Ventricular arrhythmias are a major cause of mortality in patients with ACHD. Recommendations for ICD implantation and prevention of sudden cardiac death have been recently updated.[5,28,29] Significant contributions to implantation techniques, programming, and follow-up have been published in Europace over the last 25 years allowing safe and effective application of ICD therapy for transvenous and non-transvenous ICD to this fragile population.[61–64] Despite the growing evidence for ICD implantation for primary prevention of sudden cardiac death in postoperative TOF patients, experience in patients with dextro-transposition of the great arteries (d-TGAs) after an atrial switch procedure is still sparse.[65–67] A promising new score for life-threatening ventricular arrhythmias and sudden cardiac death in these patients has recently been published.[68]

Results and side effects of ICD therapy in patients with ACHD deserve special attention. In a 2017 Europace article, Santharam *et al.*[69] analysed outcome of 42 patients with ACHD undergoing ICD implantation at a mean age of 45 years in a large referral centre. A total of 50% had repaired TOF, while 12% had surgery for d-TGA. During follow-up, 6 patients had an appropriate shock for VT/VF and 19 (45%) suffered significant complications: inappropriate shocks ($n = 11$), inappropriate anti-tachycardia pacing resulting in ventricular fibrillation ($n = 1$), infection requiring extraction ($n = 3$), lead abnormalities ($n = 3$), and pneumothorax ($n = 1$). Equal proportions of primary and secondary prevention patients received appropriate shocks. There was a 2.9% annual appropriate shock rate. However, there was a high incidence of complications with >1/3 suffering a major complication.

Lead management

Lead management remains an important issue for children and young patients with CHD. Updated consensus statements on lead management have been published recently.[5,70,71] The most common indications for lead removal in children and patients with CHD remain lead failure, venous occlusion, infection, and outgrowth of lead size. Paediatric patients are more likely to outlive the functionality of their leads, highlighting the impact of lead durability, longevity of venous access, and long-term risk of lead dysfunction.[5] There remain several knowledge gaps concerning lead management in children and patients with CHD. Data published in Europace revealed success rates of lead removal, reaching 90% with low complication rates.[72,73] Up to now, most data were published as single-centre experience with a wide age range of patients treated, while long-term data on the fate of abandoned vs. extracted leads in the young is lacking.

Subcutaneous implantable cardioverter defibrillator

The subcutaneous ICD (S-ICD) has the potential to overcome problems related to leads and vascular/anatomic abnormalities in the young and in patients with CHD.[74,75] Publications in Europace have expanded our knowledge of this technology in this population. No significant differences were observed between eligibility for an S-ICD in patients with CHD when compared with controls.[76] The multicentre European Pediatric and Young Adults with CHD Patients Registry as published just recently in Europace 2023 demonstrated good S-ICD efficacy and safety in 81 young patients. Newer implantation techniques and BMI >20 showed better outcome.[77]

Sudden cardiac arrest/death

Sudden cardiac death (SCD) is the most feared event in structural and genetic heart disease (GHD). While incredible developments have been made in the past decades to understand the genetics of cardiac channelopathies and cardiomyopathies, risk stratification remains imperfect for all conditions associated with SCD. The most robust risk calculators are those developed for hypertrophic cardiomyopathy (HCM). The European Society of Cardiology guidelines recommend implantation of a primary prevention ICD in children with two or more risk factors.[78] However, Norrish *et al.*[79] found that the positive predictive value of this approach was low in a study of 411 paediatric patients with HCM.

This important work suggests that the threshold for implanting ICDs in paediatric patients with HCM may be too low. While there are a few risk calculators for HCM, in contrast, identifying those adults with CHD at risk for SCD remains difficult. This is largely due to the diversity of the population. This is a starting point to gather data. Koyak *et al.*[80] capitalized on 3 large databases with over 25 000 patients with ACHD. They focused on echocardiographic measures of ventricular function and electrocardiographic markers. They reported that QRS duration and ventricular dysfunction progressed over time. The rate of progression of ventricular dysfunction may serve as a predictor of SCD in ACHD.

Despite the advances in understanding risk factors for SCD, the largest group of all series of SCD in the young are the unexplained or idiopathic cases. Prior to the recognition of distinct clinical entities, such as Brugada syndrome, catecholaminergic polymorphic ventricular tachycardia, and long QT syndrome, all sudden cardiac arrest (SCA) survivors with ventricular fibrillation (VF) and apparently structurally normal hearts were labelled as idiopathic VF (IVF). Over the last three decades, the definition of IVF has changed substantially, mostly as a result of the identification of the spectrum of SCA-predisposing GHDs, and the molecular evidence, by post-mortem genetic analysis (also known as the molecular autopsy), of cardiac channelopathies as the pathogenic basis for up to 35% of unexplained cases of SCD in the young. The evolution of the definition of IVF over time has led to a progressively greater awareness of the need for an extensive diagnostic assessment in unexplained SCA survivors. Nevertheless, GHDs are still underdiagnosed among SCA survivors, due to the underuse of pharmacological challenges (i.e. sodium channel blocker test), misrecognition of electrocardiogram (ECG) abnormalities/patterns (i.e. early repolarization pattern or exercise-induced ventricular bigeminy) or errors in the measurement of ECG parameters (e.g. corrected QT interval, QTc). In this review, we discuss the epidemiology, diagnostic approaches, and the controversies related to the role of the genetic background in unexplained SCA survivors with a default diagnosis of IVF.

When SCA occurs, the patient has the best chances of survival with prompt access to bystander CPR and an automated external defibrillator. Kiyohara *et al.*[81] reported on the outcomes of out-of-hospital cardiac arrest in 232 paediatric patients from 2008 to 2015. Favourable neurological outcomes were associated with bystander cardiopulmonary resuscitation (CPR) and external defibrillator (AED). Encouragingly, survival also improved over the course of the study. Similar findings were also reported by Mitani *et al.*[82] in 2013. Following a cardiac arrest, it is important to adhere to a systematic approach to the investigation of the cause. This should include a detailed history, including information from witnesses and potentially closed-circuit footage if the event was in a public space. Testing should include ECG, echocardiography, and exercise testing, at a minimum. Testing of first-degree relatives might reveal clues to latent inherited heart rhythm conditions. Genetic testing should be informed by the phenotype. Giudicessi *et al.*[83] reported on their findings with ultra-rare variants in the gene encoding the alpha subunit of the sodium channel SCN5A. The variants had all been classified as variants of uncertain significance. Four of the five variants identified did not affect ion channel function and were electrophysiologically indistinguishable from wild type. This study highlights the importance of genetic counselling and functional assessment.

An important question that is often raised is whether premature ventricular contractions (PVCs) in children are a cause for concern. In a study of 59 children with structurally normal hearts and frequent PVCs, outcome was universally reassuring, with no morbidity or mortality.[84]

Mapping and ablation: introduction

After its introduction in the 1980s, catheter-guided mapping and ablation of cardiac tachyarrhythmias have become first-line treatment options for various tachyarrhythmia substrates in children and in ACHD patients.[85,86] Since the late 1980s, radiofrequency (RF) current evolved as the preferred energy for catheter ablation, and cryoenergy as an alternative treatment mode became available in the 2000s and has been used since then in the paediatric and ACHD population.[87–89] With the introduction of three-dimensional electro-anatomical mapping systems (3D-EAM), mapping was facilitated and radiation exposure could be significantly reduced.[89–91] Due to body size and anatomical obstacles, vascular access for mapping and ablation may be difficult in children and in the ACHD population. This issue has been addressed by various articles published in EP Europace over the past 25 years.[92–95]

Paediatric ablation/cryoablation

Kammeraad *et al.*[96] reported their experience with catheter ablation of AVNRT guided by the LocaLisa® electro-anatomical mapping system. This article was one of the first to describe the usefulness of electro-anatomical mapping to safely ablate SVT with reduced use of fluoroscopy in paediatric patients. Another study from this early era of catheter ablation in paediatric patients reported a favourable outcome with a procedural success rate of 94% and a low complication rate of <1% for ablation of mainly AVNRT and accessory atrioventricular pathways but also of more uncommon tachyarrhythmias like focal atrial tachycardia, atrial flutter, and VT.[97]

The risk of AV block has been an issue since the advent of RF ablation, especially when targeting AVNRT or other substrates near the AV node. The work of Kantoch *et al.*[98] showed that despite normal AV conduction at the conclusion of the ablation procedure, late effects on AV conduction in children especially after ablation of AVNRT need to be anticipated, and patients should be followed on a regular basis in order to detect late impairment of AV conduction.

Cryoenergy has a higher safety profile compared with RF, while procedural success for SVT ablation was initially lower in paediatric patients.[99] Advanced techniques using cryoenergy have been developed by Drago *et al.*,[88,91,100–102] resulting in improved efficacy. Likewise, a recent large-scale European prospective multicentre trial on catheter ablation in paediatric patients did not find any disadvantages of cryoenergy with respect to procedural success and tachycardia recurrence for various tachycardia substrates.[89]

Mapping and ablation in congenital heart disease

The prevalence of atrial arrhythmias is constantly increasing within the ageing ACHD population significantly contributing to morbidity and mortality.[103,104] As drug treatment alone is of limited value, catheter ablation of atrial tachyarrhythmias is recommended[86,105] but may be technically challenging. Recurrences and/or new atrial tachycardias are not uncommon, often requiring multiple ablation procedures in order to achieve freedom from tachycardia.[106,107] The advent of 3D-EAM systems significantly added to our understanding of tachycardia mechanisms and improved mapping and ablation results, especially in atrial tachycardias (focal or reentrant) other than cavotricuspid isthmus–dependent atrial flutter. A non-contact, multielectrode array system was successfully applied for mapping of atrial tachycardia substrates and for activation mapping in the early era of 3D-EAM in a cohort of subjects with CHD.[108] Subsequently, non-contact mapping has almost completely been replaced by contact mapping systems using focal, mostly quadripolar catheters for mapping and ablation and later multielectrode catheters for high-density mapping. A simplified approach for 3D mapping and ablation of atrial tachyarrhythmias in patients with

ACHD using only one or two intracardiac catheters was published by Drago *et al.*,[109] yielding 90% success. This simplified approach has been adopted by many operators.

Intracardiac echocardiography

Before the availability of high-density mapping and the widespread use of contact force sensing catheters, the value of intracardiac echocardiography (ICE) in addition to 3D-EAM was used in addition to 3D-EAM to identify anatomic obstacles and to verify tissue contact in a case series of subjects with complex CHD.[110] Additionally, ICE was used in this study to aid baffle puncture in order to gain access to the pulmonary venous atrium. Procedural results were excellent and recurrences were low during mid-term follow-up, but this approach was associated with considerably increased cost.

Coherent mapping

Most recently, Klehs *et al.*[111] in a single-centre study reported on a new algorithm to facilitate the identification of critical isthmuses of IART in subjects with CHD. Results were promising making coherent contact mapping a new tool to improve and facilitate the mapping of atrial tachycardias.

Catheter ablation of atrial tachyarrhythmias in various forms of congenital heart disease

Recently, a series of studies on the outcome of catheter ablation of atrial tachyarrhythmias in various forms of CHD have been published.[112–116] A European multicentre initiative on long-term results after ablation of postoperative atrial tachyarrhythmias in subjects with TOF showed promising results and most target tachycardias were cavo-tricuspid isthmus (CTI) dependent or related to right atrial atriotomy scars. The authors suggested to extend right atrial atriotomy sutures to the inferior vena cava and to perform intraoperative CTI ablation.[112] In contrast to subjects after TOF repair, atrial tachyarrhythmias in patients after surgical closure of ASD were mostly related to atriotomy scars, while CTI-dependent atrial flutter was rare.[114] It is of note that when compared with more complex cardiac lesions, recurrences were not negligible.

Subjects after atrial switch procedure for d-TGA are at exceptionally high risk for the development of atrial tachyarrhythmias during long-term follow-up due to the presence of extensive intraatrial suture lines. Wu *et al.*[113] reported promising procedural results, although recurrences requiring a second procedure were frequent. In this study, remote magnetic catheter navigation was superior to manual catheter steering with respect to fluoroscopy time, but this technology has lost attraction when baffle puncture is performed (see below). Finally, in Ebstein's anomaly patients, tachycardia recurrences occurred in a relevant number after atrial tachyarrhythmia ablation.[116]

Vascular access

In a significant number of patients with CHD and atrial tachyarrhythmias, vascular access to the heart is limited due to vessel occlusion or due to anatomical or surgical obstacles like intraatrial baffles after atrial switch procedures for d-TGA or after Fontan-type procedures.[86] Obtaining access to the pulmonary venous atrium in patients with a Fontan circulation and after an atrial switch procedure has been outlined in two independent articles published in EP Europace. Uhm *et al.*[95] described an elegant stepwise technique in patients with a lateral tunnel or an extracardiac conduit Fontan using a standard Brockenbrough needle, a Brockenbrough needle with a snare catheter or a radiofrequency empowered transseptal needle. Krause *et al.*[94] demonstrated access to the pulmonary venous atrium in subjects after the atrial switch procedure. Both articles provide excellent guidance on access to the pulmonary venous atrium even in patients with a rigid Gore-Tex® prosthesis. Transseptal puncture may also be safely performed in smaller children, as reported by Yoshida *et al.*[93] in a cohort of subjects with a median body weight of 21.5 kg.

Last but not least, transhepatic access to the right and left atrium, respectively, has been described to be feasible and safe in a series of patients with ACHD.[92]

Summary and conclusion

This review article summarizes how EP Europace has contributed to advance the field of paediatric and congenital electrophysiology over the last 25 years. EP Europace is certainly one of the leading journals in the field of paediatric and congenital electrophysiology. In order to keep pace and even increase the impact of EP Europace, researchers and scientists are encouraged to submit their work in the future to the editors for publication in our journal.

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Data availability

All links and identifiers of the data provided are present in the references.

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