Advances in pediatric electrophysiology

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Purpose of review

The pediatric electrophysiology literature during the past year has addressed several topics that are particularly relevant for children and other patients with congenital heart disease. This paper reviews selected studies germane to physicians and health care personnel who treat pediatric and adult congenital heart patients with arrhythmias and electrophysiologic disorders.

Recent findings

Advances in arrhythmia diagnostics have been reported in pediatrics using loop monitoring, both external and implanted. Diagnostic criteria and risk stratification strategies have been refined for the congenital and inherited rhythm disorders such as cardiomyopathies and long QT syndrome. The use of therapeutic procedures such as catheter ablation for complex arrhythmias in congenital heart disease is discussed. Finally, a summary of articles on implanted devices in pediatrics and congenital heart disease is reviewed, including implantable defibrillators, atrial antitachycardia pacemakers, and cardiac resynchronization therapy in pediatrics.

Summary

Pediatric electrophysiology is a rapidly changing field, with advances seen in diagnostic evaluation of arrhythmia, refinement of risk-stratification testing, and therapeutic options such as catheter ablation and cardiac rhythm management devices. The evolution of pediatric electrophysiology from a diagnostic specialty into a therapeutic and interventional subspecialty has advanced the treatment options for children with cardiac arrhythmias and conduction disorders.

Keywords

pediatric arrhythmias, congenital heart disease, pacemaker, ablation, defibrillators

Curr Opin Pediatr 16:494–499.

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Current Opinion in Pediatrics 2004, 16:494–499

Abbreviations

HCM	hypertrophic cardiomyopathy
IART	intraatrial reentrant tachycardia
ICD	implantable cardioverter-defibrillator
LQTS	long QT syndrome
NSVT	nonsustained ventricular tachycardia
QTc	corrected QT interval
SVT	supraventricular tachycardia
TTMs	transtelephonic electrocardiographic event monitors

1040-8703

Introduction

There have been many advances in the field of cardiac electrophysiology, many of which are particularly relevant for children and patients with congenital heart disease. The pediatric electrophysiology literature during the past year has addressed several topics of interest to those who care for patients with congenital heart disease. This paper reviews selected studies of particular relevance to pediatric patients with arrhythmias and electrophysiologic disorders.

Diagnostic evaluation of the electrophysiology patient

Diagnostic evaluation of electrophysiologic disorders may be aided by various tools including monitoring devices and imaging modalities.

Cardiac event monitoring

Pediatric patients with cardiac arrhythmias may elude specific rhythm diagnosis because of the infrequent, episodic nature of their symptoms. These symptoms may include chest pain, palpitations, syncope, and presyncope. In these situations, transtelephonic electrocardiographic event monitors (TTMs) may yield documentation of the arrhythmia because they are portable and patient-activated. Whereas the utility of TTMs has been demonstrated in adults, there have been no large pediatric studies to evaluate their efficacy in children. Saarel *et al.* [1] recently published a report of 495 symptomatic pediatric patients for whom TTMs were prescribed. Patients were monitored for a mean of 103 days. Approximately half the patients did not transmit any events. Of the remaining patients, 15% had supraventricular tachycardia (SVT) documented, 91% diagnosed within 16 weeks. Sensitivity was 83% for SVT diagnosis. The negative predictive value of TTMs was 96% at 4 weeks and 98% at 16 weeks. No other significant arrhythmia besides SVT was identified, and no patient without palpitations had a significant arrhythmia; that is, no patient with chest pain or syncope without palpitations was found to have an arrhythmia [1].

Insertable cardiac monitors

When patients have worrisome though infrequent symptoms, TTMs may not yield a symptom-rhythm correlation. Insertable loop recorders implanted subcutaneously

allow continuous rhythm monitoring that is stored either when manually activated by a patient/parent or automatically when high or low rate parameters are met. Rossano *et al.* [2•] retrospectively reviewed data from three pediatric hospitals regarding the efficacy of insertable loop recorders in patients 25 years old and younger. Among 21 patients, 10 had QT prolongation, structural heart disease, or family history of sudden death. An additional 11 patients had normal hearts with palpitations, syncope/near-syncope, or acute life-threatening events. No patient experienced complications of device placement. Of 14 patients with episodes, 9 had documented arrhythmia. The authors concluded that insertable loop recorders have value in correlating arrhythmia with symptoms when noninvasive means do not make a diagnosis [2•].

Magnetic resonance imaging

Technologic advances in cardiac MRI have occurred recently, indicating that it may play a role in cardiac catheterization and ablation. Razavi *et al.* [3•] described 16 cases in which cardiac MRI was used alone or in combination with fluoroscopy. Fluoroscopy was required during some interventional and ablation cases in which catheters/devices were not compatible with MRI. The authors cite specific advantages of MRI over radiography, including better soft tissue three-dimensional visualization and cine imaging to quantify blood flow. All patients in the study received significantly lower doses of radiation than did control participants [3•]. With the development of compatible catheters and devices, MRI will be a useful tool in minimizing x-ray exposure, particularly in patients whose congenital heart disease necessitates numerous catheterization procedures.

Arrhythmias and sudden death in cardiomyopathies

Arrhythmias and sudden death may occur in hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, and congenital long QT syndrome (LQTS).

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is an inherited disease characterized by left ventricular hypertrophy and risk of ventricular arrhythmias and sudden death. The annual incidence of sudden death is 1 to 2% overall, with higher risk in young patients [4]. Implantable cardioverter-defibrillator (ICD) therapy has been successful in decreasing mortality, but its efficacy has not been well defined for younger patients. Begley *et al.* [4] studied 132 patients with HCM to assess the role of ICD therapy in this population. The average age was 34 years; one third were younger than 20 years. ICDs were used as primary prevention in 64% of patients and secondary prevention (cardiac arrest or sustained ventricular tachycardia) in 36%. In the primary prevention group, 84% of patients were event free during 5 years of follow-up, compared with 64% in the secondary prevention group. Survival in both groups was 96% overall. The risk factors associated with therapeutic shocks were age younger than 20 years and older than 40 years. Interestingly, the following were not associated with an increased risk of a shock: degree of left ventricular hypertrophy or outflow tract obstruction, nonsustained ventricular tachycardia (NSVT), or inducible ventricular tachycardia. Significant complications were common, occurring in 29% of patients. They included inappropriate shocks, infection, need for cardiac transplantation, anxiety/depression, and death. The authors encourage careful evaluation of risk in each patient and assert that some may benefit from ICD therapy [4].

Implanting an ICD in a child or young adult may have even more significant complications and psychosocial impact. Alexander *et al.* [5] recently reported a relatively large series of pediatric and young adult patients (age < 30 years) with congenital heart disease and ICDs. They noted a high rate of ICD shock therapy, both appropriate and inappropriate, but also a high lead failure rate. The strongest correlate of lead failure was change in body surface area (*ie*, growing children). The most common complications in this pediatric and young adult cohort were lead failure (21%), inappropriate shocks (18%), and ICD storm (7%). The psychologic impact of having an ICD is quite significant as well. DeMaso *et al.* [6] recently published a study on the psychosocial factors and quality of life factors in children with ICDs. They described 20 young ICD patients (age 9–19 years, mean 15 years) who completed psychologic testing and quality of life surveys. Significant associations were found between anxiety, depression, family functioning, and quality of life, particularly as perceived by their parents. They also appeared to experience a greater need for social acceptance [6].

Regarding risk stratification in HCM, Monserrat *et al.* [7] investigated the association between NSVT and prognosis in young HCM patients. They studied 531 patients with a mean age of 39 ± 15 years. Approximately 20% had NSVT on Holter monitor. These patients had greater left ventricular wall thickness and left atrial size than did those without NSVT. Specifically in patients under 30 years old, those with NSVT had a lower freedom from sudden death at 5 years than did those without NSVT (78% *vs* 94%). Interestingly, this was not true for patients older than 30 years. The frequency, rate, and duration of NSVT were not associated with prognosis at any age [7]. The authors concluded that when NSVT occurs in young patients, it likely results from a "more potent arrhythmogenic substrate." They cautioned that because sudden death occurred in some patients without NSVT, other risk factors were clearly at work.

Arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular dysplasia is another cardiomyopathy seen in young patients. Corrado *et al.* [8] found a similar life-saving value of the ICD in young adults (40 ± 15 years) with arrhythmogenic right ventricular dysplasia. They observed that half of patients received appropriate ICD therapy within the first few years after implantation, but 16% had inappropriate shocks and 14% had ICD-related complications [8].

Congenital long QT syndrome

Screening electrocardiograms are usually obtained during the evaluation of syncopal episodes to evaluate, among other parameters, the corrected QT interval (QTc). A challenging issue, however, has been the frequent overlap of QTc values from both affected patients and noncarriers of channelopathy-causing mutations. Khositseth *et al.* [9] investigated the frequency of diagnostic QTc prolongation and the frequency of nondiagnostic electrocardiograms in patients younger than 21 years with syncope. A QTc longer than 470 milliseconds was considered diagnostic because the positive predictive value in long QT syndrome approaches 100% with a QTc longer than 470 milliseconds in male persons and 480 milliseconds in female persons. Electrocardiograms from 118 patients with syncope and 118 control participants without known heart disease were studied. The mean QTc was 415 milliseconds in syncopal patients and 414 milliseconds in control participants. However, 38% of patients had a QTc between 420 and 470 milliseconds, as did 31% of control participants. Only one patient, who had exertional syncope, had a diagnostic QTc of 496 milliseconds, confirmed by genotype testing. The authors concluded that exertional syncope should prompt a thorough evaluation for long QT syndrome, but they raised concern over false positive and false negative diagnoses with a screening electrocardiogram alone because of the wide range of QTc values in both normal control participants and fainters [9].

This leads to the issue of genotyping in LQTS. With more than 200 identified mutations in seven different genes responsible for LQTS, it is clearly a genetically heterogeneous disorder. Whether genotype correlates with phenotype and whether clinical management can be based on genetic testing remains to be determined. A recent study evaluated the clinical presentation of neonatal LQTS, and correlated the genotype [10]. The authors found that newborns with sinus bradycardia and LQTS more likely had one type of LQTS (called KCNQ1 or LQT1), whereas neonates with 2:1 AV block resulting from prolonged repolarization from LQTS seemed preferentially associated with LQT2 (mutations in the *HERG* gene). They noted that these newborns with 2:1 AV block from LQT2 had a worse prognosis than newborns with LQT1 and sinus bradycardia, who could be effectively treated with β-blocker medications with or without permanent pacing [10].

Genetic testing for LQTS can even be performed in the fetus, with prenatal diagnosis feasible [11]. This may allow early neonatal diagnosis and institution of effective therapy, particularly for families with identified LQTS mutations.

Etheridge *et al.* [12] reported the use of oral potassium supplementation as therapy for LQTS. They observed that supplemental dietary potassium given over the long term improved repolarization in LQTS patients with HERG mutations [12]. This work may lead to exciting genotype-specific therapies for inherited arrhythmia conditions.

Intraatrial reentry tachycardia

Intraatrial reentrant tachycardia (IART) is associated with significant morbidity and mortality for many patients with repaired congenital heart disease, particularly after extensive atrial surgery, such as Fontan and atrial switch operations, in whom the incidence of IART is as high as 50% [13,14•–16•]. Therapeutic options for IART include antiarrhythmic medications, catheter ablation, surgery, and pacing.

Atrial antitachycardia pacing

A new generation of antitachycardia pacemaker (AT500) was tested for efficacy in a multicenter study of congenital heart disease patients with IART [14•]. Specifics of the AT500 that were studied included its ability to accurately detect atrial tachycardias, its ability to successfully pace terminate the arrhythmia, and the incidence of ventricular proarrhythmia after atrial antitachycardia pacing, which limited the use of previous antitachycardia pacemakers in the past. In 14 patients with episodes of appropriately detected atrial arrhythmias, 54% of treatable tachycardias were successfully terminated by use of antitachycardia pacing. Whereas some arrhythmias were misclassified as ventricular tachycardia, no ventricular proarrhythmia occurred after antitachycardia pacing therapy. One limitation of the AT500 appears to be misclassification of atrial tachycardias with 1:1 AV conduction as ventricular in origin. Despite this, the AT500 was used safely and often effectively in this relatively small number of congenital heart disease patients [14•].

Catheter ablation and surgical revision

Much attention has been focused on the roles of catheter and surgical ablation for treatment of IART in patients with palliated congenital heart disease. Despite reports of high acute success rates with catheter ablation, arrhythmia recurrence continues to be problematic in this population. Kannankeril *et al.* [16•] attempted to identify acute and late recurrence rates after acutely successful radiofrequency ablation of IART. Diagnoses included primarily single ventricle with Fontan palliation and transposition of the great arteries with Mustard or Senning repair. Acute success was reported in 93% of pa-

tients and was defined as no inducible, sustained IART. Recurrence occurred in 34% of patients, 88% within 1 year. Patients with Fontan physiology had the highest recurrence rate, although the sample size was too small to demonstrate a significant difference. The authors invoked the likelihood of inadequate lesion size and depth with conventional radiofrequency ablation catheters as the principle reason for recurrence. This study did not include the use of irrigated tip catheters or higher-powered ablation sources, which may create lesions of greater size and depth [16•].

During the past few years, much has been published regarding cryoablation of arrhythmia circuits at the time of surgical Fontan conversion for refractory atrial arrhythmias [17,18]. It has been postulated that by redirecting systemic venous flow, the resultant decrease in atrial pressure leads to a decrease in atrial size and hence a less arrhythmogenic substrate. Despite this operation, however, patients are often left with recurrent or new arrhythmias [15•]. The success of intraoperative cryoablation within the right atrium for patients with IART and/or left atrium for atrial fibrillation using preoperative and intraoperative mapping has been reported. A recent series included 10 patients who underwent cryoablation at the time of conversion of an atriopulmonary connection to an extracardiac Fontan [15•]. In addition, 9 of the 10 received a dual chamber pacemaker. The patients ranged from 12 to 33 years of age at the time of reoperation. Indications for surgery were not mutually exclusive and included refractory arrhythmias (8 patients), protein-losing enteropathy (2), systemic venous obstruction (1), and deteriorating NYHA class (all patients were class III or IV). There were 2 deaths, ultimately resulting from sepsis and multisystem organ failure. All 8 survivors showed an improvement in NYHA classification. Of the 6 surviving patients with a history of arrhythmia, 4 were noninducible during electrophysiologic testing 2 months postoperatively; 2 had inducible nonsustained IART, and 1 had clinical recurrence of paroxysmal IART [15•].

A second study by Deal *et al.* [17] evaluated their experience with arrhythmia surgery, specifically excluding patients who underwent Fontan conversion. Of 29 patients with symptomatic, medically refractory arrhythmias, 27 had structural heart disease repaired during the same surgical procedure. The patients ranged from 6 days to 45 years of age. Arrhythmia mechanisms included atrial reentry tachycardia, atrial fibrillation, automatic atrial tachycardia, AV reciprocating tachycardia, AV node reentry tachycardia, and ventricular tachycardia. Medical therapy had been unsuccessful in all patients; 11 catheter ablation procedures had been performed in 7 patients. The surgical techniques for arrhythmia elimination included endocardial and/or epicardial resection, isolation, and cryoablation as appropriate for the specific arrhythmia. The Cox maze III procedure was used to treat atrial fibrillation. Postoperative electrophysiologic studies were performed before hospital discharge on the surviving patients with a history of reentrant arrhythmia mechanisms [17]. Surgical mortality was 7%. Four patients had early postoperative tachycardia. During follow-up electrophysiologic study, supraventricular tachycardia was inducible in 1 of 14 patients, and ventricular tachycardia was inducible in 2 of 5 patients, who underwent ICD implantation. During a median follow-up time of 47 months, 2 additional patients had a clinical recurrence of SVT, and 2 required pacemaker implantation for late sinus node dysfunction. AV block did not develop in any patient. The authors concluded that the surgical approach to arrhythmia management may be successfully combined with repair of congenital heart disease, and that this approach may be of particular benefit to patients who are not good candidates for catheter ablation therapy or in whom this therapy has been unsuccessful, such as neonates and individuals with complex anatomy [17].

Prophylactic atrial incisions for the prevention of late IART have been evaluated in the short term [19]. Spontaneous IART was uncommon, but the interventional atrial incisions altered atrial conduction time and did not increase morbidity [19]. Longer follow-up times will be necessary to determine whether these prophylactic atrial cryoablation lesions or other acute surgical interventions will be beneficial for IART prevention.

Electrical cardiac resynchronization therapy

Adult patients with dilated cardiomyopathy or heart failure have been shown to benefit from biventricular pacing (or cardiac resynchronization therapy) with regard to cardiac index, VO_{2max}, and quality of life. Bundle branch block or interventricular conduction delay often accompanies heart failure as well as some forms of congenital heart disease, either preoperatively or postoperatively, and may result in ventricular dysfunction caused by asynchronous myocardial contraction [20•]. Biventricular pacing is an attempt to resynchronize ventricular contraction by pacing both ventricles, thereby improving overall ventricular function. In patients with left bundle branch block, cardiac resynchronization therapy counteracts the underlying electrical and mechanical dyssynchrony, leading to improved contractility, function, exercise tolerance, and quality of life [21–23].

Cardiac resynchronization in pediatrics and congenital heart disease

Extrapolating the potential benefits of cardiac resynchronization therapy in adults with heart failure to pediatric patients with structural cardiac disease, Zimmerman *et al.* [20•] studied the effects of multisite ventricular pacing in patients undergoing surgery for congenital heart disease. Twenty-nine patients aged 1 week to 17 years were

categorized into three groups: single-ventricle anatomy, two-ventricle anatomy undergoing ventricular surgery, and two-ventricle anatomy undergoing another type of cardiac surgery. All patients had preexisting bundle branch block/interventricular conduction delay or were expected to have this postoperatively. None required pacing for sinus bradycardia or heart block. Two atrial and three ventricular epicardial leads were placed at the time of surgery, with the ventricular leads placed as far from one another as possible. Atrial synchronous ventricular pacing was established postoperatively, and the AV interval was adjusted to yield the narrowest QRS complex while simultaneously pacing two ventricular sites. Multisite pacing resulted in significant narrowing of the QRS complex, improved cardiac index, and increased systolic blood pressure overall. The authors assert that multisite pacing after surgery for congenital heart disease in children may facilitate weaning from inotropic agents, thereby limiting side effects such as increasing myocardial oxygen consumption [20•]. A limitation cited by the authors is that there was no control group that underwent single-site pacing.

Janousek *et al.* [24] reported using biventricular pacing in infants with dilated cardiomyopathy associated with pacing in complete heart block. They described two infants with severe dilated cardiomyopathy after 3 to 4 years of dual chamber pacing for complete AV block (congenital in one patient, postsurgical in one). Remarkably, after only 1 month of biventricular pacing, there was improved left ventricular function and reverse remodeling. This suggests that biventricular pacing may be beneficial in infants with cardiomyopathy associated with complete AV block and conventional right-sided pacing [24].

Roofthooft *et al.* [25] also reported the improvement in a 2-week-old neonate who underwent congenital heart surgery and experienced complete AV block, for which she had placement of a dual-chamber pacemaker, with epicardial leads sutured on the right atrium and right ventricle. She subsequently experienced left ventricular failure with an ejection fraction of 25%. The pacing system was upgraded to biventricular, with placement of a left ventricular epicardial lead, and she had marked clinical improvement [25].

Right ventricular resynchronization in congenital heart disease patients

As above, the effects of biventricular pacing in left ventricular failure have been studied. Dubin *et al.* [26•] recently published a study of the effects of electrical resynchronization on right ventricular dysfunction in congenital heart disease. Right bundle branch block is a common outcome after surgery for congenital heart lesions. Right ventricular pressure and/or volume loading may also be present, with attendant right ventricular enlargement and dyskinesis. The authors studied seven patients with congenital heart disease, right bundle branch block, and right ventricular dysfunction. Data obtained during cardiac catheterization included QRS duration, cardiac index, and right ventricular dP/dt_{max}. These parameters were measured during sinus rhythm, asynchronous atrial pacing, and dual chamber pacing with an AV delay of 90% of the PR interval. The QRS duration decreased in all patients during dual chamber pacing, whereas dP/dt_{max} increased with dual chamber pacing but not with asynchronous atrial pacing, by a mean of *22%*. Cardiac index also increased during resynchronization with dual chamber pacing in 6 of 7 patients. The pacing site yielding the narrowest QRS duration also improved cardiac index the most in 6 of 7 patients, but was not associated with the site producing the best dP/dt_{max}. Although the role of AV delay was not evaluated, and the number of patients was small, this study demonstrated the acute benefits of right ventricular resynchronization in a select group of congenital heart disease patients [26•].

Conversely, Stephenson *et al.* [27] recently demonstrated that pacing the right ventricle from standard apical sites without resynchronization may actually lengthen depolarization time. They measured QRS duration from patients with tetralogy of Fallot and ICDs, and noted longer QRS durations with right ventricular pacing compared with intrinsic conduction in patients with right bundle branch block. The QRS duration could be shortened acutely in a subgroup of patients by optimization of the AV interval, potentially allowing right ventricular resynchronization in these patients [27]. Whether long-term right ventricular resynchronization will be feasible and effective using standard ICD leads remains to be determined.

Conclusion

Pediatric electrophysiology is a rapidly changing field, with advances seen in diagnostic evaluation of the arrhythmia patient, refinement of risk-stratification testing, and therapeutic options such as catheter ablation and cardiac rhythm management devices.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- • Of special interest
- •• Of outstanding interest

1 Saarel EV, Stefanelli CB, Fischbach PS, et al.: Transtelephonic electrocardiographic monitors for evaluation of children and adolescents with suspected arrhythmias. Pediatrics 2004, 113:248–251.

2 • Rossano J, Bloemers B, Sreeram N, et al.: Efficacy of implantable loop recorders in establishing symptom-rhythm correlation in young patients with syncope and palpitations. Pediatrics 2003, 112:e228–e233.

Implantable loop monitors provided symptom-rhythm correlation in patients with syncope, near-syncope, and palpitations, with and without structural heart disease after noninvasive means failed.

3 • Razavi R, Hill DL, Keevil SF, et al.: Cardiac catheterization guided by MRI in children and adults with congenital heart disease. Lancet 2003, 372:1877–1882.

MRI was used in combination with fluoroscopy to decrease radiation exposure and improve soft tissue visualization during cardiac catheterization of congenital heart disease patients.

4 Begley DA, Mohiddin SA, Tripodi D, et al.: Efficacy of implantable cardioverter defibrillator therapy for primary and secondary prevention of sudden cardiac death in hypertrophic cardiomyopathy. Pacing Clin Electrophysiol 2003, 26:1887–1896.

5 Alexander ME, Cecchin F, Walsh EP, et al.: Implications of implantable cardioverter defibrillator therapy in congenital heart disease and pediatrics. J Cardiovasc Electrophysiol 2004, 15:72–76.

6 DeMaso DR, Lauretti A, Spieth L, et al.: Psychosocial factors and quality of life in children and adolescents with implantable cardioverter-defibrillators. Am J Cardiol 2004, 93:582–587.

7 Monserrat L, Elliott PM, Gimeno JR, et al.: Non-sustained ventricular tachycardia in hypertrophic cardiomyopathy: an independent marker of sudden death risk in young patients. J Am Coll Cardiol 2003, 42:873–879.

8 Corrado D, Leoni L, Link MS, et al.: Implantable cardioverter-defibrillator therapy for prevention of sudden death in patients with arrhythmogenic right ventricular cardiomyopathy/dysplasia. Circulation 2003, 108:3084–3091.

9 Khositseth A, Martinez MW, Driscoll DJ, et al.: Syncope in children and adolescents and the congenital long QT syndrome. Am J Cardiol 2003, 92:746–749.

10 Lupoglazoff JM, Denjoy I, Villain E, et al.: Long QT syndrome in neonates: conduction disorders associated with HERG mutations and sinus bradycardia with CKNQ1 mutations. J Am Coll Cardiol 2004, 43:826–830.

11 Tester DJ, McCormack J, Ackerman MJ: Prenatal molecular genetic diagnosis of congenital long QT syndrome by strategic genotyping. Am J Cardiol 2004, 93:788–791.

12 Etheridge SP, Compton SJ, Tristani-Firouzi M, et al.: A new oral therapy for long QT syndrome long-term oral potassium improves repolarization in patients with HERG mutations. J Am Coll Cardiol 2003, 42:1777–1782.

13 Dilawar M, Bradley SM, Saul JP, et al.: Sinus node dysfunction after intra-atrial lateral tunnel and extracardiac conduit Fontan procedures. Pediatr Cardiol 2003, 24:284–288.

14 • Stephenson EA, Casavant D, Tuzi J, et al.: ATTEST investigators: Efficacy of atrial antitachycardia pacing using the Medtronic AT500 pacemaker in patients with congenital heart disease. Am J Cardiol 2003, 92:871–876.
Atrial arrhythmia detection and antitachycardia pacing efficacy of the Medtronic AT500 pacemaker was evaluated in 28 patients with congenital heart disease and atrial arrhythmias. Antitachycardia pacing was successful in converting 54% of treatable episodes.

15 • Weinstein S, Cua C, Chan D, et al.: Outcome of symptomatic patients undergoing extracardiac Fontan conversion and cryoablation. J Thorac Cardiovasc Surg 2003, 126:529–536.
Ten patients undergoing Fontan revision surgery had empiric cryoablation and pacemaker implantation during the operation in an attempt to treat refractory atrial arrhythmias. Seven patients currently require no antiarrhythmic agents. Surgical mortality was 20%.

16 • Kannankeril PJ, Anderson ME, Rottman JN, et al.: Frequency of late recurrence of intra-atrial reentry tachycardia after radiofrequency catheter ablation in patients with congenital heart disease. Am J Cardiol 2003, 92:879–881.
Catheter ablation was performed in 40 patients with congenital heart disease and intraatrial reentry tachycardia. The acute success rate was 87%. Arrhythmia recurred in 34%.

17 Deal BJ, Mavroudis C, Backer CL: Beyond Fontan conversion: surgical therapy of arrhythmias including patients with associated complex congenital heart disease. Ann Thorac Surg 2003, 76:542–553; discussion 553–554.

18 Petko M, Myung RJ, Wernovsky G, et al.: Surgical reinterventions following the Fontan procedure. Eur J Cardiothorac Surg 2003, 42:185–196.

19 Collins KK, Rhee EK, Delucca JM, et al.: Modification to the Fontan procedure for the prophylaxis of intra-atrial reentrant tachycardia: short-term results of a prospective randomized blinded trial. J Thorac Cardiovasc Surg 2004, 127:721–729.

20 • Zimmerman FJ, Starr JP, Koenig PR, et al.: Acute hemodynamic benefit of multisite ventricular pacing after congenital heart surgery. Ann Thorac Surg 2003, 75:1775–1780.
Twenty-nine patients with congenital heart disease and a wide QRS complex underwent multisite ventricular pacing for cardiac resynchronization. Multisite pacing led to improved cardiac index and increased systolic blood pressure.

21 Bristow MR, Saxon LA, Boehmer J, et al.: Cardiac resynchronization therapy with or without an implantable defibrillator in advanced chronic heart failure. N Engl J Med 2004, 350:2140–2150.

22 Auricchio A: Beyond the measurement of QRS complex toward mechanical dyssynchrony: cardiac resynchronisation therapy in heart failure patients with normal QRS duration. Heart 2004, 90:479–481.

23 Turner MS, Bleasdale RA, Mumford CE, et al.: Left ventricular pacing improves haemodynamic variables in patients with heart failure with a normal QRS duration. Heart 2004, 90:502–505.

24 Janousek J, Tomek V, Chaloupecky V, et al.: Dilated cardiomyopathy associated with dual-chamber pacing in infants: improvement through either left ventricular cardiac resynchronization or programming the pacemaker off allowing intrinsic normal conduction. J Cardiovasc Electrophysiol 2004, 15:470–474.

25 Roofthooft MT, Blom NA, Rijilaarsdam ME, et al.: Resynchronization therapy after congenital heart surgery to improve left ventricular function. Pacing Clin Electrophysiol 2003, 26:2042–2044.

26 • Dubin AM, Feinstein JA, Reddy VM, et al.: Electrical resynchronization: a novel therapy for the failing right ventricle. Circulation 2003, 107:2287–2289.
Seven patients with right ventricular dysfunction and right bundle branch block had improved RV performance during atrioventricular pacing with RV resynchronization.

27 Stephenson EA, Cecchin F, Alexander ME, et al.: Relation of right ventricular pacing in tetralogy of Fallot to electrical resynchronization. Am J Cardiol 2004, 93:1449–1452.