sciendo Journal of Cardiovascular Emergencies 2018;4(4):197-200
DOI: 10.2478/jce-2018-0027

CLINICAL UPDATE

Modern Technology for Prevention of Sudden Cardiac Death – a Clinical Update on Device Therapy in Children with Congenital Heart Diseases

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ABSTRACT

Sudden cardiac death in children is one of the most devastating conditions that can be encountered in acute cardiac care. Intracardiac device therapy, providing prompt and effective treatment in malignant ventricular arrhythmia or in severe conduction abnormalities, is a promising tool to reduce the incidence of this fatal condition. However, the implementation of device-based therapy in the pediatric population is currently limited by the lack of clinical studies on large number of subjects. As a result, indications for device therapy in pediatric patients are still unclear in many circumstances. There are also several particularities related to device implantation in pediatric age, such as the somatic growth leading to a mismatch between chamber size and lead length, or the difficulties of implantation technique in children with small body weight. This study aims to present an update on the current advantages and limitations of device-based therapy for treating severe malignant arrhythmia or conduction disorders in children at risk for sudden cardiac death.

Keywords: congenital heart disease, sudden cardiac death, intracardiac defibrillators

ARTICLE HISTORY

Received: October 30, 2018
Accepted: November 28, 2018

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BACKGROUND

Device therapy has a well-established role in the treatment of various heart diseases in the adult population. Intracardiac pacemakers are widely used for treating atrioventricular conduction disturbances, while intracardiac defibrillators (ICD) are first-line therapies for treating severe arrhythmia and preventing sudden cardiac death. In parallel, resynchronization devices have been validated as efficient tools for treating heart failure in selected cases with electrical dyssynchrony, restoring contraction synchronism by simultaneous stimulation of the left and right ventricle. While the role of device therapy at adult age is widely accepted nowadays, many controversies still exist regarding the role of implantable devices in children with congenital heart diseases (CHD). One of the main reasons for this is the lack of large clinical trials in a pediatric population and the significant heterogeneity of this group.[1] Recent technological developments have made possible the manufacturing of small sized devices for pediatric patients. However, in the current era of evidence-based medicine, there are only several clinical studies proving the effectiveness of device-based therapy in CHD. As a result, indications for device therapy in pediatric patients are still unclear in many circumstances.[2]

SUDDEN CARDIAC DEATH IN CHILDHOOD

Sudden cardiac death (SCD) is the most severe type of cardiovascular emergency that can be encountered in childhood.

In children, the incidence of SCD has been reported to be approximately 1/100,000 individuals, being usually encountered in pediatric patients with various forms of ventricular tachycardia (VT) or presenting postoperative arrhythmia after surgical repair of CHD.[3] Other frequently identified causes include cardiomyopathies, myocarditis, coronary artery abnormalities, or conduction system anomalies.[3]

Sudden cardiac death in childhood occurs also in association with CHD. In the CHD group, there are several specific diseases that are associated with a higher risk for severe ventricular arrhythmia and SCD. One of these high-risk subgroups is represented by children with CHD and inherited channelopaties.[4] Other diseases associated more often with a higher arrhythmic risk are the tetralogy of Fallot and the transposition of the great arteries (TGA).[5] After surgical treatment for tetralogy of Fallot, the incidence of ventricular arrhythmia has been reported to be as high as 1.2% to 3.0% per decade.[6] TGA is associated with a higher risk for SCD, which can result from malignant ventricular arrhythmia or rapid ventricular response to atrial fibrillation, and also with an increased frequency of abnormalities of the conduction system, which have been reported in as much as 25% of children with TGA.[6,7] Patients with CHD surviving to adult age present also a relatively high incidence of ventricular arrhythmia, approximately 0.1% to 0.2% per year, and implantation of an ICD device in these patients can be life-saving.[8]

In many cases, a genetic predisposition exposes the children to a higher risk of various heart diseases. For instance, angiotensin gene polymorphism has been identified in secondary pulmonary arterial hypertension in children with congenital heart disease.[9] This kind of genetic inheritance has been described in several types of malignant arrhythmia and SCD in pediatric patients. Genetic testing and counseling may have a significant impact on the decision-making process with respect to indicating ICD therapy to children from families carrying an increased risk of SCD. Another exemplification of the utility of genetic tests in CHD is in patients with tetralogy of Fallot, in whom severe ventricular arrhythmia may occur after surgical correction, especially in case of coexisting long QT gene mutations or polymorphism.[10]

PARTICULARITIES OF IMPLANTABLE DEFIBRILLATORS FOR THE PREVENTION OF SUDDEN CARDIAC DEATH IN A PEDIATRIC POPULATION

Several studies reported an age-related suboptimal performance of ICD leads in children with CHD and implanted ICD.[11] One particular problem associated with ICD in pediatric patients is related to the high frequency of inappropriate shock delivery, which has been reported more frequently in pediatric patients than in adults, varying between 19% and 46% according to different authors. This condition usually requires system revision, which carries the inherent risks associated to reintervention.[12–19] The type of device used (dual-chamber or single-chamber) did not prove to influence the frequency of this complication. In children with implanted ICD, inappropriate shock is probably caused by lead displacement occurring with somatic growing or by lead malfunction resulting from a more active lifestyle.[20]

IMPLANTATION OF INTRACARDIAC DEVICES IN CHD – A CHALLENGING PROCEDURE

The complexity of cardiac anatomy in CHD can make implantation of an intracardiac device a very challenging procedure. Frequently, children with CHD have various types of intracardiac communications that make it difficult to guide the leads to the target area. Furthermore, the risk of complications following the surgical correction of CHD is not negligible, and these postoperative complications can significantly impact the evolution of the patients.[21] Device therapy in neonates or young infants is even more challenging due to their low weight. In a recent report, 34% of implants at age below 1 year were associated with lead malfunction events.[22] However, successful pacemaker implantation has been reported even in small children weighing less than 10 kg.[23] This indicates that device therapy in this age category is extremely challenging, and a proper selection of devices and implantation technique is essential for improving the success rate and to preserve the functionality of the device on a longer term.

IMPLANTABLE PACEMAKERS FOR TREATING CONDUCTION DISORDERS IN PEDIATRIC AGE

Cardiac conduction disorders can occur in structurally normal hearts, in various types of CHD, or following the surgical correction of complex structural heart diseases.[24] Atrioventricular blocks in children can result in Adam-Stokes crisis or even SCD. It has been demonstrated that the implantation of cardiac pacemakers in atrioventricular block following surgical intervention in CHD was associated with a significant decrease in the risk of SCD and episodes of Adam-Stokes attacks.[25] As for the functionality of intracardiac pacemakers in a pediatric population, a retrospective study on 663 pediatric patients with CHD who

underwent pacemaker implantation reported functional epicardial pacing at 10 years after implant in more than 60% of patients.[26]

One issue related to ventricular pacing in children with CHD is related to left ventricular dysfunction resulting from inappropriate ventricular pacing, especially in severe CHD such as double outlet right ventricle, TGA and ventricular septal defect, or atrioventricular canal defect.[27] However, in a small study on 42 patients, Friedberg *et al.* demonstrated that resynchronization therapy in the immediate postoperative period can improve hemodynamic status and increase the cardiac index following the surgical correction of CHD.[28] This shows that an efficient pacing could improve hemodynamic status in children with severe heart diseases and should be judiciously indicated in selected cases.

SUBCUTANEOUS AND LEADLESS IMPLANTABLE DEVICES IN CHILDREN

Subcutaneous ICDs have emerged as a viable alternative to devices implanted via transvenous route, reducing the risks related to the implantation procedure (risk of cardiac tamponade, pneumothorax, hemothorax, arterial puncture, or hematoma), as well as the longer term risk of lead dysfunction.[29,30] Especially in children, who are still at growing age, a mismatch between the size of the lead and the size of the growing body is inherent and could be avoided by the implantation of subcutaneous devices.[30] This mismatch could determine lead displacement requiring reintervention and placement of a new, longer lead.[23] Subcutaneous implantable devices could avoid this risk of lead mismatch and represent a promising alternative to traditional ICDs, especially in pediatric patients.

Leadless pacemakers have also been proposed as alternatives to traditional pacemakers starting with 2012. These are self-contained devices delivered to the right ventricle, avoiding the need for creating a subcutaneous pocket and the need for transvenous lead as well.[7] However, the effectiveness of this new type of therapy in children has not been proved so far.

CONCLUSION

In conclusion, device therapy can improve survival and quality of life in children at risk for SCD, by treating severe arrhythmia or conduction disorders that can have devastating consequences. In certain conditions, the implantation of a proper device can be extremely challenging, especially in infants with small weight or in the case of complex CHD with difficult anatomy. This therapy could be life-saving, providing urgent and effective treatment in several forms of major cardiovascular emergencies; however, larger studies are required to validate the most effective type of device or implantation technique in the complex and heterogenous group of children with CHD

CONFLICT OF INTEREST

Nothing to declare.

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