

## Outcomes of long-term endocavitary cardiac pacing in children

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### Abstract

**Background:** The development of neonatal cardiology and cardiac surgery, as well as the earlier diagnosis of congenital defects, arrhythmias, and cardiac conduction disorders, contributed to the systematic increase of the number of children and young adults undergoing electrophysiological procedures, as well as to the reduction of the age at which these procedures are performed. Children requiring permanent cardiac pacing constitute 1–2% of the entire population qualified for cardiac pacemaker implantation.

**The aim of this study** was to present the results of permanent pacing therapy employed in children undergoing endocavitary pacemaker implantation in long-term follow-up.

**Material and methods:** The study group included 106 children up to 12 years after pacemaker implantation. The age range of the children was 2–17 years, while their weight was between 13 and 58.8 kg. The group included 91 children after surgical repair of congenital heart defects, 7 patients with congenital complete atrioventricular block, 4 patients with postinflammatory sinus node dysfunction, and 4 patients with symptomatic bradycardia after heart transplantation.

**Results:** All 106 patients were implanted with endocardial system types consistent with their indications for pacing. The mean duration of the procedure was 92 min, while the average duration of fluoroscopy was 9 min 37 s. The average battery lifespan in follow-up was 5 years.

**Conclusions:** Transvenous pacemaker implantation in children is a minimally invasive method, feasible for use even in small children.

**Key words:** endocardial pacing, cardiac pacing in children.

### Streszczenie

**Wstęp:** Dzięki rozwojowi kardiologii i kardiochirurgii okresu noworodkowego, wczesnej diagnostyce wad wrodzonych oraz zaburzeń rytmu i przewodnictwa serca systematycznie obniża się wiek i zwiększa liczba dzieci i młodych dorosłych poddawanych procedurom elektrofizjologicznym. Dzieci wymagające leczenia stałą stymulacją serca stanowią 1–2% całej populacji kwalifikowanej do implantacji kardiostymulatora.

**Celem pracy** było przedstawienie wyników leczenia stałą stymulacją serca u dzieci poddanych endokawitarnej implantacji kardiostymulatora w obserwacji długoterminowej.

**Materiał i metody:** Obserwacji poddano grupę 106 dzieci do 12 lat po implantacji. Zakres wieku dzieci wynosił 2–17 lat, waga 13–58,8 kg. Wśród badanych było 91 dzieci z blokiem przedsionkowo-komorowym lub dysfunkcją węzła zatokowego po chirurgicznej korekcji wrodzonej wady serca, 7 pacjentów z wrodzonym całkowitym blokiem przedsionkowo-komorowym, 4 pacjentów z pozapalną niewydolnością węzła zatokowego, 4 pacjentów z objawową bradykardią po transplantacji serca.

**Wyniki:** U wszystkich 106 pacjentów implantowano układ endokawitarny z rodzajem stymulacji zgodnym ze wskazaniami. Średni czas zabiegu wynosił 92 min, średni czas fluoroskopii 9 min 37 s. W obserwacji odległej średni czas żywotności baterii wynosił 5 lat.

**Wnioski:** Przewidywalna implantacja układu stymulującego jest metodą małoinwazyjną i możliwą do zastosowania u dzieci.

**Słowa kluczowe:** endokawitarna stymulacja, stymulacja serca u dzieci.

### Introduction

The dynamic development of neonatal cardiology and cardiac surgery, as well as the significant improvement of

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maternity care with diagnostic support, including well-developed fetal echocardiography, has contributed to the early diagnosis of congenital defects, arrhythmias, and cardiac conduction disorders. The number of children and young adults undergoing electrophysiological procedures is growing, while the age at which these procedures are performed is decreasing. According to data reported in the USA, children requiring permanent cardiac pacing constitute 1-2% of the entire population qualified for cardiac pacemaker implantation [1]. Currently, two methods of cardiac pacemaker implantation are used in pediatric patients requiring permanent pacing. The first method, widely used during the early years of electrical pacing in children, is the epicardial method. It utilizes epicardial electrodes sutured to the myocardium using a transdiaphragmatic approach; the pacemaker battery is placed in the epigastrium, under the left costal margin. Epicardial electrodes are sometimes applied during surgical heart defect repair. The selection of epicardial pacemaker implantation in a pediatric population may be necessary due to anatomical conditions preventing the use of the transvenous method. The drawbacks of this method are its high invasiveness (even with the use of thoracoscopic techniques that are rarely employed in small children) and shorter lifetime of the system resulting from the decreasing parameters of epicardial pacing and leading to additional interventions [2]. The transvenous method constitutes an alternative surgical implantation technique. It consists in accessing the myocardium via the central vascular system – by performing a puncture of the subclavian vein or venesection of the cephalic vein. The pacemaker pouch is then located under the pectoral fascia or the pectoral muscle [3]. Endocavitary implantation is less invasive and does not necessitate a long-lasting immobilization of the child as part of the postoperative regime. The anxiety related to the use of this method usually concerns the technical problems associated with the implantation in small children, potential future interventions

accompanying the linear growth of the child, and the inactive electrodes left in the cardiovascular system resulting from the many years of cardiac pacing.

The selection of the pacing method must be based on the individual analysis of the child's anatomy and body mass, the presence of proper cardiovascular connections, as well as the experience of the center in which the pacemaker implantation is to be performed. It is important to emphasize that pacemaker implantation is performed increasingly more often in very young children, which leads to many years of subsequent cardiac pacing. Therefore, when choosing the technique for implantation, not only the technical aspects of the procedure should be taken into consideration, but also the long-term effects of the treatment. Few studies concerning the analysis of the above-mentioned problems have been conducted so far. This work presents our single-center experience with long-term transvenous cardiac pacing in children.

**Aim of the study**

The aim of this work is to assess the effectiveness of heart pacing and its clinical consequences in the long-term follow-up of children undergoing pacemaker implantation using the endocavitary method.

**Material and methods**

A group of 106 children who had pacemakers implanted using the transvenous method during the period between January 1999 and the end of June 2012 was included in a retrospective 12-year follow-up. The age range of the children was 2-17 years, median 12 years. Weight: 13-58.8 kg, median: 38.8 kg. There were 34 patients weighing 20 kg or less, which constituted 32% of the studied group. The group did not include any children weighing less than 10 kg. The most common recommendations for pacemaker implantation in the study group were atrioventricular conduction disorders

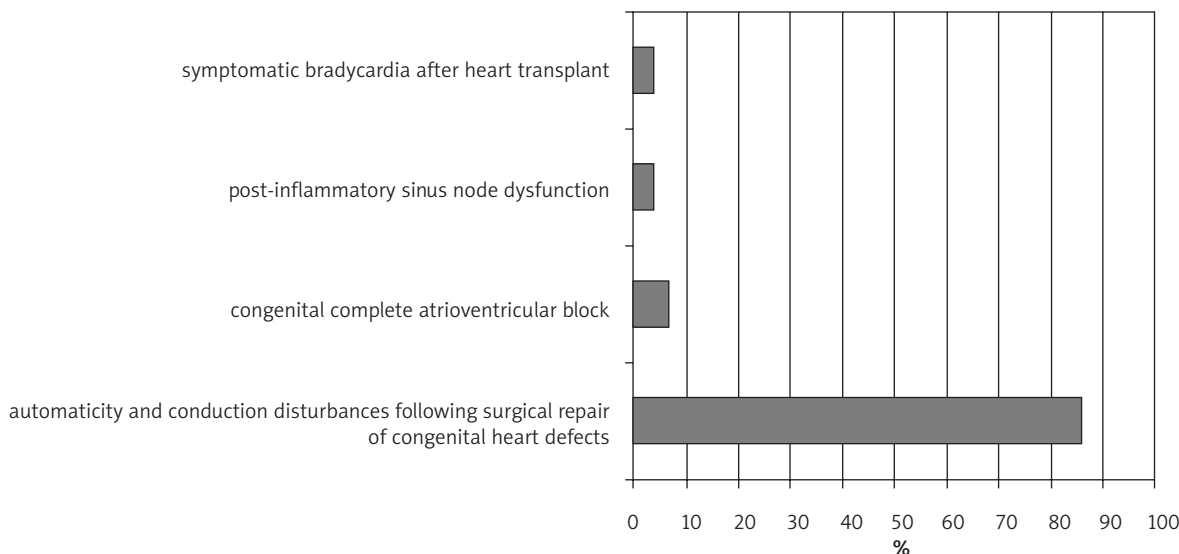


Fig. 1. Indications for pacemaker implantation in this population

or sinus node dysfunction resulting from the surgical repair of congenital heart defects (91 patients). Other indications included complete congenital atrioventricular block (7 patients), post-inflammatory sinus node insufficiency with normal myocardial anatomy (4 patients), and bradycardia after a heart transplant (4 patients). The percentage distribution of recommendation types is presented in Fig. 1. Qualification for the procedure and postoperative care took place at the SUM Child Cardiology Ward of the Department of Cardiology, Congenital Heart Defects, and Electrotherapy of the Silesian Center for Heart Diseases in Zabrze. The procedures were conducted by the team of the Electrophysiology and Heart Pacing Laboratory at the mentioned clinic.

The pacemakers were implanted via venous access, by means of cephalic vein venesection in 57 patients, while in the remaining 49 patients, it was necessary to perform subclavian vein puncture. The following pacemakers were implanted: 84 DDDR, 17 AAIR, 5 WIR, and 1 VDD. Ventricular electrodes were placed in the apex of the right ventricle, leaving a loop in the atrium, which allowed for the "straightening" of the electrode as the child grew. Active fixation was used in 19 patients, while passive fixation was employed in the remaining cases. Atrial electrodes were fixed near the atrial auricle. Active atrial electrodes were used in 51 patients; passive electrodes were utilized in the remaining cases. As in adult patients, the pacemaker was implanted opposite to the dominant limb. The pacemaker pouch was located in subcutaneous tissue. Medtronic, Biotronic, and St. Jude pacemakers were implanted. In children over 16, the procedure was performed using very precise local anesthesia; in all the younger children, it was performed in deep sedation supplemented by local anesthesia. First check-up of the pacemakers took place one week after the implantation. Subsequently, after one month, optimal pacing parameters were programmed for each patient. Further check-ups were routinely performed every 6 months. If the child's linear growth amounted to 10 cm, the location of the electrodes was assessed by means of X-ray imaging. In 25 patients, a telemonitoring system was applied: Home-monitoring Biotronik or CareLink Medtronic, depending on the device type.

## Results

Among all the children qualified for endocavitary pacemaker implantation, the procedure was abandoned in one case because of the difficulties in the preparation of the cephalic vein and the unsuccessful attempts to perform subclavian vein puncture. In all the remaining 106 patients, the endocavitary pacemakers with appropriate pacing types were successfully implanted. The average duration of the procedure was 92 min, while the average duration of fluoroscopy was 9 min 37 s. The follow-up included the immediate postoperative period and the period of up to 12 years after implantation. In 17 children (16% of the study group), the follow-up period exceeded 10 years. The observed complications included: 2 early dislocations of atrial electrodes (2 and 8 days after surgery) and 2 dislocations of ventricular

electrodes (6 days and 1.5 months after implantation). An 11-year-old girl after dual-chamber pacemaker implantation experienced impairment of the venous circulation in the left forearm and palm. Heparin and acenocoumarol were used in the therapy, and the symptoms subsided completely. A 16-year-old girl required pacemaker repositioning under the pectoral muscle 6 months after the implantation due to pacemaker migration. In a 17-year-old boy after a heart transplant, infective endocarditis was diagnosed 2 years after pacemaker implantation, which necessitated the reimplantation of the pacemaker. None of the patients in whom venous access was achieved by means of subclavian vein puncture suffered from pneumothorax. In long-term follow-up, it was necessary to replace the pacemaker battery 5 years after the procedure on average. Only 4 children required the implantation of a new ventricular electrode due to the loss of proper pacemaker sensing and increased excitability threshold. In all these cases, attempts were made to perform transvenous extraction of the inactive ventricular electrode, which was successful in 3 out of 4 patients. Three children had their ventricular electrodes "extended" 2 years on average after the procedure, by first preparing the pacemaker and then pushing the ventricular electrode with a director. This helped delay the necessity of implanting additional ventricular electrodes in these children. In an 8-year-old girl, 5 years after the implantation, the ventricular and atrial electrodes were damaged; both electrodes were removed, and a new endocavitary DDDR system was implanted. One 11-year-old patient required an upgrade of his DDD pacemaker to the CRT-P version. The pacing parameters at every follow-up stage are presented in Table I.

## Discussion

Among the pediatric patients qualified for cardiac pacemaker implantation, the most numerous group is formed by children in whom automaticity and conduction disorders were caused by surgical repair of congenital heart defects. Congenital heart defects, understood as structural anomalies in the circulatory system, occur in 0.18-1.9% of neonates born alive. Half of these are complex defects burdened with high mortality and requiring cardiac intervention during the first year of the child's life [4]. The beginning of treating congenital heart defects was in the late 1930s. In 1938 in Boston, Robert Gross closed a patent ductus arteriosus in a 7.5-year-old girl. In 1944, a Swedish surgeon, Clarence Crafoord, performed the first aortic coarctation in the world [5]. Another person who had an enormous impact on the development of pediatric heart surgery and electrophysiology was Ake Senning, a Swedish cardiac surgeon. In 1958 in Stockholm, Ake Senning performed a surgical inversion of ventricular inflow in a child with a reverse arrangement of ventricular outflow (transposition of arterial trunks), correcting the defect physiologically. After the procedure, the ventricular inflow and outflow were reversed in comparison to the normal arrangement. This method was used for many years in children with such defects, until anatomical repair of transposed arterial trunks became wide-

**Tab. I.** Pacing parameters at various follow-up stages

	Intraoperative measurements	1 month after implantation	12 months after implantation	36 months after implantation	Over 5 years after implantation
RV sensing (mV)	3.5-30.0	4.0-30.0	4.0-30.0	4.0-30.0	4.0-30.0
RA sensing (mV)	1.8-6.5	2.0-4.5	2.2-4.5	2.0-5.0	2.2-5.5
RV impedance ( $\Omega$ )	225-1600	250-1500	220-1500	220-1800	218-1800
RA impedance ( $\Omega$ )	280-1700	250-1500	342-1600	318-960	313-1040
RV pacing threshold (V/ms)	0.5-1.5*	0.4-2.5*	0.4-2.5*	0.5-1.5*	0.5-3.5*
RA pacing threshold (V/ms)	0.5-2.0*	0.4-2.0*	0.4-2.0*	0.5-2.0*	0.5-2.0*

\*With pulse width of 0.4 ms

spread. In the same year (1958), Ake Senning performed the first cardiac pacemaker implantation in the world in a patient with postinflammatory atrioventricular block [6]. Implantation of cardiac pacemakers in children began in the 1960s. In 1962, a pacemaker was implanted in a 7-year-old boy with postoperative atrioventricular block after tetralogy of Fallot repair. The first reports concerning the use of permanent pacing in neonates come from 1966. In Poland, the first pacemaker implantation in a child, resulting from third-degree atrioventricular block secondary to a heart tumor, was performed by Professor Bjork in 1965 [7]. Currently, over 3000 surgical repair procedures of congenital heart defects are performed in Poland each year [8]. The prevalence of complete postoperative heart block in pediatric patients after congenital heart defect repair is estimated as 1-5.8% [9, 10]. Complete postoperative heart block sometimes subsides on its own; however, when it continues for more than 7 days after surgery, it constitutes an indication for pacemaker implantation (recommended: ACC/AHA/NASPE). When it comes to prevalence, the second most common indication for the use of permanent pacing in children is sinus node dysfunction after the repair of such congenital heart defects as atrial septal defect, partial and complete anomalous pulmonary venous drainage, complex heart defects corrected using the Fontan method, as well as the atrial repair of transposition of the great vessels [11]. Sinus node dysfunction which is not a complication of heart defect repair performed in the atria may be caused by hypervagotonia, inflammation, metabolic disorders, ion and hormonal imbalance, or drugs. It may also coexist with anorexia in young individuals [12]. In patients after heart transplants, the transplanted hearts are devoid of vagus nerve fibers and sympathetic terminals, which may result in an increased basal heart rate. These patients may sometimes experience sinus bradyarrhythmia or atrioventricular nodal rhythm, which necessitates pacemaker implantation due to heart rate insufficiency [13]. Indications for pacemaker implantation in children with bradyarrhythmia after heart transplantation occur rarely, which may also be related to the relatively low number of pediatric patients undergoing heart transplants. The prevalence of complete congenital atrioventricular block ranges from 1: 15 000 to 1: 22 000 in live-born neonates. In 17-39% of

these children, the disease coexists with other pathologies of the circulatory system. In the majority of cases, these are: atrioventricular discordance, transposition of the great arteries, and significant interventricular septal defects [12]. Connective tissue disease of the mother is often listed as one of the etiological reasons for third-degree congenital heart block. It has been revealed that the SS-A/Ro or SS-B/La antibodies present in the mother's organism pass through her placenta and damage the conduction system of the already formed heart [14]. In an American single-center study, including 105 seropositive mothers and 107 neonates live-born from these mothers, as many as 67 children (63%) required pacemaker implantation before the age of 1 [15]. Children requiring pacemaker implantation are likely to need this method of treatment for the rest of their lives. In such cases, the method is often used for decades and every subsequent intervention involves additional risk of complications. At the initial stage of permanent cardiac pacing in children, all implantations were performed using the epicardial method. It turned out, however, that epicardial pacemakers pose numerous problems in long-term follow-up. Tissue fibrosis and scarring around the epicardial electrode caused an increase in the excitability threshold, necessitating the programming of a higher amplitude, which resulted in decreased lifetime of the pacemaker's battery. The use of epicardial implantation was also more often associated with exit block - an excitability threshold too high for pacing to be possible, which requires the implantation of a new pacemaker. Despite the increasingly common use of epicardial electrodes releasing steroidal drugs, some authors claim that the average time of the correct functioning of epicardial pacemakers is only 4 years [2, 16]. The application of epicardial electrodes onto the myocardium after opening the pericardial sac bears the risk of post-pericardiotomy syndrome and increases the likelihood of perioperative complications resulting from the immobilization of the patient.

The transvenous method of pacemaker implantation has been widely used since 1958, when the endocavitary electrode was first introduced for clinical use by Seymour Furman, an American cardiologist [17]. The method consists in accessing the myocardium via the central vascular system. Electrodes are introduced through the subclavian vein



and the superior vena cava to the right atrium or further, through the tricuspid orifice, into the right ventricle, or, in the case of resynchronization pacing, to the left ventricle through the coronary sinus system. In comparison to epicardial electrodes, endocavitary electrodes are characterized by longer lifespan, resulting from lower risk of damage and lower pacing thresholds. This method is undoubtedly less invasive than epicardial implantation, which is widely employed in adult patients. Endocavitary electrode implantation in older children with anatomically healthy hearts is not different from the same procedure performed in adults. Technical problems may arise when the weight of the child is very low or if the child underwent surgical congenital heart defect repair. The majority of congenital heart defect repair procedures are performed via the right atrium and the tricuspid valve, which is the reason for the presence of scarring in the atrial region; the auricle of the right ventricle is often excised [18]. In the study group, such children were implanted with atrial electrodes with active fixation. It is also important to mention that, in children with transposition of the great arteries repaired using the Senning or Mustard procedures, it was more difficult to attain effective pacing because of the small existing area of normal atrial tissue. Low body mass of the child and the resulting small vein size may cause difficulty in achieving vascular access for transvenous electrode implantation. It may also be associated with higher risk of thromboembolic complications than in adult patients. Within the studied group of children, circulatory disorders occurred in one 11-year-old girl, immediately after the procedure. The ailments subsided after the administration of heparin and an oral anticoagulant. According to some authors, subclavian vein puncture should be avoided in children because of the higher risk of pneumothorax in comparison to adult patients [3, 19]. In the presented material, none of the patients in whom venous access was achieved by means of subclavian vein puncture suffered from pneumothorax. Infection of the pacing system is the most serious complication of implantation, with its mortality rate reaching 24% in adult patients in long-term follow-up [20]. Unsuccessful treatment of infection within the area of the tissue pouch of the pacemaker results in the appearance of bacterial vegetation on the endocardial electrode, which may cause endocarditis and lead to the child's death due to sepsis. In a publication by Cohen concerning pacemaker infections in children, the prevalence of superficial and deep pacemaker infections in a follow-up group of 267 patients was 4.9% and 2.3%, respectively. In a multifactorial analysis of the same author, the most significant predisposing factors were trisomy 21 and reoperations [21]. In the studied group of children, infective endocarditis was diagnosed 2 years after pacemaker implantation in a 17-year-old boy after a heart transplant. The whole pacing system was removed; after successful treatment, a new pacemaker was implanted on the opposite side. It is also important to mention that antibiotic therapy is routinely performed at our center before the procedure, as a method for preventing pace-

maker infections. Because of their higher physical activity, children are more often susceptible to mechanical damage to pacemaker electrodes or batteries. Damage to atrial and ventricular electrodes occurred in an 8-year-old girl, 5 years after cardiac pacemaker implantation. After the transvenous removal of the damaged electrodes, a new pacing system was implanted. Since the expected duration of permanent pacing in children is counted in decades, long-term endocavitary pacing of the right ventricular apex may raise doubts when it comes to the early choice of transvenous implantation. This site is frequently selected because of the technical ease of applying the electrodes, ensuring significant stability and adequate pacing parameters. This non-physiological direction of myocardial activation leads to paradoxical movement of the interventricular septum, increased left ventricular pressure, and, in consequence, to the remodeling of the heart and left ventricular dysfunction. The prevalence of post-pacing damage to the left ventricle in children is approximately 7% [22]. In the studied group, an 11-year-old boy was diagnosed with dilated cardiomyopathy 4 years after the implantation of a VVI system, which resulted in the need for resynchronization pacing. A new, significant problem, which concerns not only children but all patients undergoing long-lasting endocavitary electrotherapy, is the prolonged presence of endocavitary electrodes in the cardiovascular system, particularly when inactive electrodes are left behind after implanting new ones due to the child's growth or after changing the pacing mode. Inactive electrodes left in the vascular system increase the risk of pressure ulcers in the pacemaker pouch or the subclavian area. They may also disrupt the venous circulation and increase the risk of vein obstruction by irritating the vessel walls. Furthermore, they raise the likelihood of thrombosis, pulmonary embolism, and tricuspid valve dysfunction. Moreover, they may cause damage to the insulation of an active electrode, resulting from friction between electrodes in the cardiovascular system [23, 24]. In the discussed material, inactive ventricular electrodes were removed in 3 cases, damaged atrial and ventricular electrodes were removed in the case of an 8-year-old girl, and a DDD system was removed from a 17-year-old patient due to infection. It needs to be pointed out that the benefits of the removal have to outweigh the risk related to the procedure. Therefore, each case needs to be analyzed individually, taking into consideration all indications, the experience of the center, and the patient's examination results.

## Conclusions

A 12-year-long follow-up is a relatively short period of time in the long-lasting electrotherapy predicted for the discussed group of patients. However, the analysis of the presented results indicates the low invasiveness of transvenous pacemaker implantation, which may be used even in young children and in difficult anatomical conditions caused by heart defects or their surgical repair. In long-term follow-up, it is associated with a low risk of unfavorable clinical consequences resulting from this method of pacing. The studied

group of patients may experience the following problems: inadequacy of pacing parameters resulting from electrode dislocation caused by the child's linear growth, the necessity of replacing inactive electrodes, and the fact that every change of battery requires an additional intervention, burdened with increasingly higher risk of complications, including the most serious ones, such as infection of the pacing system.

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