*Lidia Ziółkowska, Monika Kowalczyk, Katarzyna Bieganowska, Wanda Kawalec

Current therapeutic options in children with hypertrophic cardiomyopathy – own experience**

Aktualne metody terapeutyczne u dzieci z kardiomiopatią przerostową – doświadczenia własne

Department of Pediatric Cardiology, The Children's Memorial Health Institute, Warszawa
Head of Department: prof. Wanda Kawalec, MD, PhD

Key words

hypertrophic cardiomyopathy, sudden cardiac death, cardioverter-defibrillator, septal myectomy, heart transplant, children

Słowa kluczowe

kardiomiopatia przerostowa, nagły zgon sercowy, kardiowerter-defibrylator, myectomia, przeszczep serca, dzieci

Summary

Introduction. The clinical course of hypertrophic cardiomyopathy (HCM) in children is very heterogeneous. In some children, the thickened heart muscle causes progressive heart failure and life-threatening arrhythmias. HCM is the most common cause of sudden cardiac death.

Aim. The aim of study was retrospective analysis of clinical profile, current therapeutic options and sudden cardiac death risk assessment in children with HCM.

Material and methods. We analyzed 108 children, mean age 9.7 ± 5.46 yrs with HCM diagnosed in the years 1991-2013. Mean follow-up was 6.4 ± 4.83 yrs. Patients demographics, clinical symptoms, treatment strategy as well as the results of echocardiography, ECG, 24 h Holter ECG, exercise test were analyzed.

Results. Of the 108 patients studied 77 (71%) were treated only pharmacologically, in 4 (3.7) children RF catheter ablation was done, in 8 (7.4%) patients septal myectomy was performed. In 17 (15.7%) children with risk factors for sudden cardiac death ICD for primary (12/17) and secondary prevention (5/17) was implanted. The heart transplant was qualified in 6 (5.5%) patients who had symptoms of progressive heart failure. Among 108 patients 11 (10.6%) children died. The mean annual mortality rate was 1.34.

Conclusions. In most children with HCM lifestyle modification and appropriate pharmacological therapy are sufficient, additional medical interventions are not necessary. In patients with symptomatic left ventricular outflow tract obstruction refractory to medical therapy a surgical myectomy should be considered. High-risk patients ought to be prospectively identified and ICD implantation should be considered, however further research is required to establish better criteria for primary prevention in children.

Streszczenie

Wstęp. Przebieg kliniczny kardiomiopatii przerostowej (HCM) u dzieci jest bardzo różnorodny. U niektórych dzieci, przerost mięśnia sercowego powoduje postępującą niewydolność serca i występowanie zagrażających życiu zaburzeń rytmu serca. HCM jest najczęstszą przyczyną nagłej śmierci sercowej.

Cel pracy. Celem pracy była retrospektywna analiza spektrum klinicznego, aktualnych metod terapeutycznych i stratyfikacji ryzyka nagłego zgonu sercowego u dzieci z HCM.

Materiał i metody. Analizą objęto 108 dzieci, w wieku średnio 9,7 ± 5,46 lat z HCM rozpoznaną w latach 1991-2013. Średni okres obserwacji wynosił 6,4 ± 4,83 lat. Analizowano dane demograficzne pacjentów, ich objawy kliniczne, metody leczenia, jak również wyniki badania echokardiograficznego, EKG, 24-godzinnego EKG metodą Holtera oraz testu wysiłkowego.

Wyniki. Spośród 108 analizowanych pacjentów u 77 (71%) stosowano tylko leczenie farmakologiczne, u 4 (3,7) dzieci wykonano ablację RF, u 8 (7,4%) pacjentów wykonano operacyjne wycięcie mięśnia przegrody międzykomorowej. U 17 (15,7%) dzieci z czynnikami ryzyka nagłej śmierci sercowej kardiowerter-defibrylator został wszczepiony w prewencji pierwotnej u 12 dzieci i w prewencji wtórnej u 5. Do przeszczepu serca zakwalifikowano 6 (5,5%) chorych, u których występowały objawy postępującej niewydolności serca. Spośród 108 pacjentów 11 (10,6%) dzieci zmarło. Średnia roczna śmiertelność wynosiła 1,34.

Address/adres:

*Lidia Ziółkowska
Department of Pediatric Cardiology
The Children's Memorial Health Institute
Al. Dzieci Polskich 20, 04-730 Warszawa
tel. + 48 (22) 815-73-70
fax + 48 (22) 815-73-65
lidia@wa.home.pl

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Wnioski. U większości dzieci z HCM modyfikacja stylu życia i odpowiednia terapia farmakologiczna są wystarczające, dodatkowe interwencje medyczne nie są konieczne. U pacjentów z objawowym zawężaniem drogi odpływu lewej komory, opornym na leczenie farmakologiczne, należy rozważyć leczenie chirurgiczne. Pacjenci wysokiego ryzyka, powinni być prospektywnie identyfikowani i wszczepienie ICD powinno być brane pod uwagę, jednak konieczne są dalsze badania w celu ustalenia lepszych kryteriów kwalifikacji do prewencji pierwotnej u dzieci z kardiomiopatią przerostową.

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a relatively common genetic cardiac disease (0.47 cases per 100.000 children per year) that is heterogeneous with respect to disease-causing mutations, clinical presentation, prognosis, and treatment strategies. HCM is a myocardial disease characterized by hypertrophy of the left ventricle which is not secondary to congenital heart disease or arterial hypertension. The severity of cardiac hypertrophy, etiology, as well as the clinical course of HCM in children is varied, resulting in a large spectrum of clinical and phenotypic expression (1, 2). Left ventricular hypertrophy is usually asymmetrical. Most are occupied septum and anterolateral free wall of the left ventricle, concentric hypertrophy occurs frequently and occasionally apex hypertrophy (3). Hypertrophic obstructive cardiomyopathy was diagnosed in 25% of patients with HCM, they have an obstruction and systolic pressure gradient in the left ventricular outflow tract (LVOT). In a subgroup of children with HCM, the thickened heart muscle can cause signs and symptoms, such as shortness of breath, fatigue, syncope, chest pain, progressive heart failure and problems in the heart's electrical system resulting in life-threatening arrhythmias (3, 4). Hypertrophic cardiomyopathy is the most common cause of sudden cardiac death (SCD) in the young (including competitive athletes) (5-7). A subgroup of patients with hypertrophic obstructive cardiomyopathy (HOCM) remain severely symptomatic despite optimal medical therapy. Septal myectomy reduces or eliminates left ventricular outflow obstruction and produces marked symptomatic improvement (8, 9).

Treatment strategies include drug treatment for exertional dyspnea (β-blockers, verapamil), pharmacological treatment and radiofrequency catheter (RF) ablation for arrhythmias and the septal myectomy operation, which is the standard of care for severe refractory symptoms associated with marked outflow obstruction. The alcohol septal ablation and pacing are alternatives to surgery for selected adult patients. High-risk patients may be effectively protected against sudden cardiac death with the implantable cardioverter-defibrillator (ICD). In children with progressive heart failure pharmacological treatment of heart failure is used, and in the absence of improvement they are qualified for a heart transplant.

AIM

Retrospective analysis of clinical profile, current therapeutic options and SCD risk assessment in children with HCM hospitalized in the Department of Pediatric Cardiology in the years 1991-2013. Mean follow-up was 6.4 ± 4.83 yrs (ranged from 5 months to 20 yrs).

MATERIAL AND METHODS

We analyzed 108 patients, 44 girls, 64 boys, mean age 9.7 ± 5.46 years (ranged from 4 month to 17.7 yrs) with hypertrophic cardiomyopathy. Patients demographics, clinical symptoms (shortness of breath, fatigability, syncope, pre-syncope, chest pain, heart failure, arrhythmias), family history of HCM and SCD, treatment strategy as well as the results of echocardiography, chest X-ray, 12-leads ECG, 24h Holter ECG, exercise test with assessment of blood pressure response to exercise were analyzed. HCM was diagnosed in the presence of left ventricular hypertrophy (more than two SDs from the normal range corrected for body size (BSA); z-score ≥ 2) in the absence of haemodynamic conditions that could account for the observed degree of hypertrophy. Left ventricular hypertrophy as % of mean normal range relative to BSA and z-score were calculated. Family history of SCD was defined as one or more SCD in relatives < 40 years of age or SCD in a relative with confirmed HCM at any age. Unexplained syncope was defined as unexplained transient loss of consciousness at or prior to first evaluation. Vasovagal syncope was not considered a risk factor for SCD. Abnormal systolic blood pressure response to exercise (ABPRE): < 25 mmHg rise in systolic blood pressure from rest to peak exercise and/or > 10 mmHg drop from maximal systolic blood pressure when exercised to exhaustion. Nonsustained ventricular tachycardia (NSVT) was defined as ≥ 3 consecutive ventricular extra systoles at a rate of ≥ 120 beats/min lasting < 30 s during Holter monitoring. In all patients, cardiological test results and data from the family history have been analyzed regarding the presence of major risk factors for SCD according to the Expert Group of the American and the European Society of Cardiology (10-12) recommendations such as sudden cardiac arrest (SCA) in an interview, sudden cardiac death in the family history, syncope of unknown etiology, left ventricular thickness ≥ 30 mm, spontaneous sustained ventricular tachycardia, abnormal blood pressure response during exercise and episodes of NSVT in 24-hour Holter ECG recording.

RESULTS

Of the 108 patients studied 77 (71%) were treated only pharmacologically, with 4 children underwent RF catheter ablation (3.7%), in 8 (7.4%) patients septal myectomy was performed. In 17 (15.7%) children with risk factors for sudden cardiac death ICD for primary prevention (12/17) and secondary prevention (5/17) was implanted. The heart transplant was qualified in six patients (5.5%) who had symptoms of progressive heart failure, not responsive to drug treatment efficacy. Table 1 lists the baseline characteristics of the 108 patients studied.

Table 1. Baseline variables of 108 patients studied.

Clinical parameters	Study group
Cohort mean age yrs ± SD, range	9.7 ± 5.46, (0.04-17.7)
Male, n (%)	65 (60%)
Age at diagnosis mean yrs ± SD, range	6 ± 5.17 (0.01-17.1)
Diagnosis of HCM in infancy, n (%)	6 (5%)
Family history of HCM, n (%)	48 (44%)
Family history of SCD, n (%)	25 (23%)
Resuscitated sudden cardiac arrest, n (%)	5 (4.6%)
Syncope, n (%)	15 (14%)
Pre-syncope, n (%)	23 (21%)
Chest pain, n (%)	30 (28%)
Heart palpitations, n (%)	18 (17%)
Dyspnoea on exertion, n (%)	22 (20%)
Maximum ventricular wall thickness, mm, mean % of mean normal range relative to BSA z-score	 5.7-44 (15) 131-657 (242) 2.04-10.4 (3.7)
Asymmetric septal hypertrophy, n (%)	74 (68)
Concentric left ventricular hypertrophy, n (%)	31 (29%)
Apical left ventricular hypertrophy, n (%)	3 (3%)
LVOT gradient > 30 mmHg, n (%)	24 (22%)
NSVT, n (%)	15 (14%)
Beta-blockers, n (%)	68 (88%)
Calcium-blockers, n (%)	8 (10%)
Antiarrhythmics (sotalol or amiodarone), n (%)	9 (8%)
ACE-inhibitors, n (%)	9 (8%)
Diuretics (furosemide or spironolactone), n (%)	25 (23%)
Myectomy, n (%)	8 (7.4%)
RF ablation, n (%)	4 (3.7%)
ICD implantation, n (%)	17 (15.7%)
Primary prevention, n (%)	12 (11%)
Secondary prevention, n (%)	5 (4.6%)

RADIOFREQUENCY CATHETER ABLATION OF THE ACCESSORY PATHWAY

In 7 patients with electrocardiographic features of pre-excitation (Wolff-Parkinson-White syndrome)electrophysiological study was done, in 4 (3.7%) children accessory atrioventricular pathway was diagnosed and RF ablation was performed. The mean age at the ablation was 12.1 ± 5 yrs (ranged from 6.7 to 17 yrs). Maximum left ventricular wall thickness ranged from 6.6 to 18.9 mm, an average of 14 mm (from 157 to 378% of mean normal range relative to BSA, medium 240%, z-score ranged from 2.3 to 9.7, average 5.8). Of the 4 patients with accessory pathways, in 2 children episodes of supraventricular tachycardia and reentry atrioventricular tachycardia were present. RF ablation was effective in 2 patients with an accessory atriventricular pathways located in left upper wall, after procedure no evidence of pre-excitation was found. One patient underwent RF ablation of two accessory atrioventricular pathways located in the septal posterior wall and right lateral wall. It was a good early effect of ablation-no conduction by bundle of Kent, but perhaps there is a His-Purkinje conduction (a real Mahaim fiber)because after ablation in electrocardiography features of pre-excitation persisted. It was concluded that the early result of RF ablation is good, remote requires further observation. In one patient RF ablation of two accessory atrioventricular pathways located in the left bottom and lateral wall was performed. After intervention, no signs of preexitation in electrocardiography was observed. During ventricular pacing narrow complexes – the earliest retrograde activation of the His bundle around the left side occured. Due to the location (indicated transseptal access), duration of application abandoned to continue the procedure. This patient received amiodarone and is expected to repeat RF ablation. All of these four patients after RF ablation receive beta-blockers.

SEPTAL MYECTOMY

Septal myectomy operation was performed in 8 (7.4%) children with HOCM with a maximum systolic pressure gradient in LVOT ≥ 50 mmHg at rest and clinical symptoms refractory to medical therapy. The mean age at diagnosis of HCM was 1.5 years (ranged from 1 month to 4 years), with 5 (63%) patient HCM diagnosed in infancy. The mean age at operation was 10.2 ± 6.5 yrs (ranged from 11 months to 15.9 yrs). In 7 (88%) patients asymmetric septum hypertrophy and in one child concentric hypertrophy of the left ventricle was diagnosed. Maximum septal thickness ranged from 16 to 40 mm, an average of 25 mm (from 258 to 620% of mean normal range relative to BSA, medium 389%), z-score ranged from 4.0 to 9.7, average 5.9). Preoperatively, mean maximum systolic gradient in LVOT was 98 mmHg, ranged from 50 mmHg to 150 mmHg. Transaortic extended left ventricular septal myectomy was performed in all patients with no early deaths. Mean maximum systolic gradient in LVOT decreased from 98 to 38 mmHg (ranged from 16 mmHg to 120 mmHg). Postoperatively, in one patient a residual LVOT gradient 120 mmHg (before surgery 150 mmHg)was present. After the operation, in all patients improvement in exercise tolerance (NYHA functional class I or II), reduced dyspnea at rest were observed. Changes in LVOT gradient before and after septal myectomy are shown in figure 1.

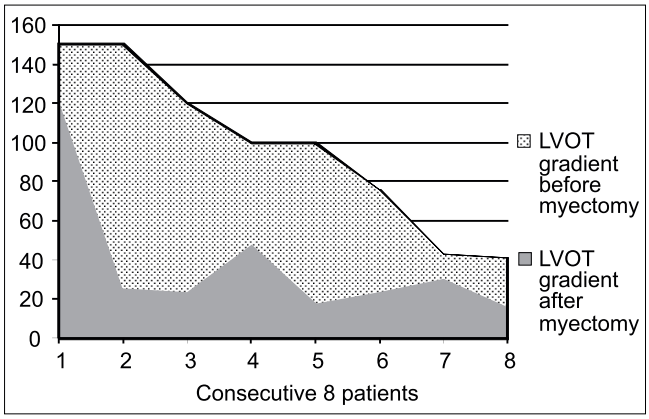

Fig. 1. Change in LVOT gradient after septal myectomy.

During the mean follow-up period of 3.9 years (ranged from 1 month to 12 years) the maximum systolic gradient in LVOT ranged from 4 to 130 mmHg, an average of 67 mmHg. In four patients reccurence of obstructionin LVOT (maximum systolic gradient > 50 mmHg) was observed (one patient died suddenly 10 years after surgery), and in the remaining four children gradient maintained at a value lower than before the operation.

PRIMARY AND SECONDARY PREVENTION OF SUDDEN CARDIAC DEATH

Of the 108 analyzed patients, in 17 (15.7%) children with major risk factors for SCD ICD for primary (12/17) and secondary prevention (5/17) was implant ed. The mean age at diagnosis of HCM was 7.4years (ranged from 1 month to 15.2 years), with 4 (24%) children HCM diagnosed in infancy. The mean age of the patients at the time of ICD implantation was 13.7 ± 3.9 years (from 12 months to 16.8 years). Maximum left ventricular wall thickness ranged from 7 to 42 mm, an average of 25 mm (from 157 to 620% of mean normal range relative to BSA, medium 360%, z-score ranged from 2.3 to 9.7, average 5.5). In the group of 5 patients in whom an ICD was implanted as secondary prevention of SCD were the major SCD risk factors, such as: sudden cardiac arrest successfully resuscitated in all 5 children, sudden cardiac death due to HCM in a family history in 4 patients, syncope in 3 children, thickness of the interventricular septum ≥ 30 mm in 1 patient, NSVT episodes in 2 patients, abnormal blood pressure response during exercise in 2 patients. In these patients, appropriate ICD discharge occurred in 3 (60%) children, inappropriate in one (20%) child because of damage to the electrode. In the group of 12 patients who underwent ICD implantation for primary prevention of SCD were the major risk factors, such as sudden cardiac death due to HCM in a family history in 5 (42%) patients, syncope in 5 (42%) children, the thickness of the left ventricular ≥ 30 mm in 4 (33%) patients, NSVT episodes in 3 (25%), abnormal blood pressure response during exercise in 2 (20%) patients. In 5 patients coexistence of two major risk factors for SCD were present: sudden cardiac death in the family due to HCM and NSVT (n = 1), syncope and left ventricular thickness ≥ 30 mm (n = 1), episodes of NSVT and the thickness of the left ventricular ≥ 30 mm (n = 1), sudden death due to HCM in the family and syncope (n = 1), NSVT after exercise and ABPR during exertion (n = 1). In 1 patient three major risk factors for SCD were found: sudden cardiac death due to HCM in the family, left ventricular thickness ≥ 30 mm, and ABPR during effort. In 6 children the presence of one of the major risk factor for SCD was found: syncope (n = 3), sudden cardiac death due to HCM in the family (n = 2), massive hypertrophy of the left ventricle ≥ 30 mm (n = 1). In these group of patients appropriate ICD discharges occurred in 1 (8.3%) child, and inadequate discharge in 2 (16.6%) patients(damage to the electrodes in 1 child, and displacement of the right ventricular electrode to coronary sinus in the second patient).

PROGRESSIVE HEART FAILURE

For a heart transplant six patients (5.5%) who had symptoms of progressive heart failure, not responsive to drug treatment efficacy were qualified. The mean age at diagnosis of HCM in this group was 1.9 years (ranged from 1 month to 4 years), with 2 (33%) patients HCM diagnosed in the first week of life. Maximum left ventricular wall thickness ranged from 9 to 20 mm, an average of 15 mm (from 178 to 426% of mean normal range relative to BSA, medium 285%), z-score ranged from 2.7 to 6.3, average 4.3. In all six children concentric hypertrophy of the left ventricle was found. In 3 (50%) patients furthermore hypertrophy of the right ventricle was observed. All patients in this group had symptoms of severe heart failure, 5 children were in NYHA class III, 1 in NYHA class II. In 2 (1.8%) patients heart transplantation was done (in 2 year old girl and 16 year old boy), three patients died while waiting for a transplant, one child is still on the waiting list. Among 108 patients, 11 (10.6%) children died, aged an average of nine years (from 6 months to 16 years). The mean annual mortality rate was 1.34. Sudden cardiac death occurred in 4 (3.8%) children, mean age 11.4 years (ranged from 8.3 to 13 years), but due to progressive heart failure died seven (6.7%) patients with a mean age of 7.7 years (ranged from 6 months to 16 years).

DISCUSSION

The clinical course of HCM in children is very heterogeneous and therefore the outcome is difficult to predict. Hypertrophic cardiomyopathy often goes undiagnosed, because many of those patients have few, if any, symptoms. These children often lead normal lives with no significant problems. They may not even realize that they have the condition until it is found during a routine medical exam. In our group of children, in 71% of patients used only pharmacological treatment, while in 29% it was necessary to apply a more aggressive therapy such as RF catheter ablation, septal myectomy,

ICD implantation and heart transplant. In analyzed patients accessory atrioventricular pathways were present in four (3.7%) children which is comparable with the results of other authors (5%) (13). According to own and other authors opinion (14) particular attention should be pay to the proper ECG interpretation in children with HCM. The accessory atrioventricular conduction manifested by short PR interval with or without associated delta wave can be seen. This could be due to coexistent Wolf Parkinson White syndrome seen in less than 5% of HCM patients or due to accessory atrioventricular conduction without any anatomic accessory pathway. Certain genetic diseases like storage disorders are known to phenotypically mimic HCM and are frequently associated with short PR interval or WPW syndrome. These include PRKAG2 (Protein Kinase AMP activated Gamma 2) mutation of Fabry disease or LAMP2 (Liposomal Associated Membrane Protein 2) mutation seen in Pompe and Danon diseases. It is important to differentiate these genetic variants of glycogen storage disorder as Fabry, Pompe and Danon because they are multisystem diseases with pleotropic manifestations. In these patients true preexcitation with single or multiple accessory pathways is frequently seen and patients die at a very young age secondary to life threatening ventricular arrhythmias. Considering the high morbidity and mortality in patients with LAMP2/PRKA2 mutations, genetic testing is advisable in phenotypic hypertrophic cardiomyopathy patients with short PR interval. HCM can occur in children with genetic syndromes such as Noonan syndrome or its allelic form Leopard syndrome, which is the result of a mutation of the gene PTPN11. In the analyzed group of patients, 7 (6.5%) children were confirmed by genetic testing Leopard syndrome (4 pts), Noonan (2 pts) and Costello syndrome (1 pt).

Dynamic pressure gradient in LVOT arises from the movement and adhesion of anterior mitral valve leaflet to the hypertrophied septum. In approximately 5% of children the gradient in the LVOT is caused by abnormal hypertrophy and trailers front anterior papillary muscle to the mitral leaflet, or by a massive central part of the ventricular hypertrophy. In many patients, cardiac hypertrophy coexists with mitral apparatus abnormality (enlargement and elongation leaflets) or papillary muscles (15, 16). Medical treatment is the first-line therapy for patients with symptoms due to LVOT obstruction, however, septal myectomy seems to be anexcellent treatment option for those intolerant of or unresponsive to medical therapy. It is worth emphasizing that in the study group only in 8 (7.4%) children systolic pressure gradient in LVOT $\geq$ 50 mmHg was found. Research published showed that the obstruction of LVOT was present in 40% to 59% of children, which undoubtedly stems from the fact that the HOCM group included patients with resting systolic gradient $\geq$ 16 mmHg (2, 4, 23). In our patients the mean age at septal myectomy was 10.2 years, so that, they were younger than children in the recently published work (12.9 years) and they had a greater thickness of the interventricular septum (z-score 5.9 vs 3.2) (18). Postoperatively, in all patients exercise tolerance improved and dyspnea at rest subsided, mean maximum gradient decreased from 98 mmHg to 38 mmHg. In the study group septal myectomy was performed less frequently compared with literature data (7.4% vs 24%) and smaller gradient reduction in the LVOT was obtained, whereas in none of the our patient heart block requiring pacemaker implantation occured, which is reported in 1-5% of children (2, 4, 17). It should be noted that during follow-up period (mean 3.9; maximum 12 years) in half of the operated patients recurrence of gradient in LVOT was observed. In the opinion of other authors the common causes of recurrent LVOT obstruction include inadequate myectomy at the first operation, midventricular obstruction and anomalies of mitral valve and papillary muscles as opposed to recurrent growth of muscle during the follow-up period. This is particularly noted in children who appear to be at higher risk of having such recurrence. Most often, inadequate myectomy at the initial operation is due to failure to extend the myectomy far enough toward the midventricle and apex of the heart. However, myectomy is more challenging in children because of smaller structures (aortic root) and, consequently, incomplete myectomy or aortic or mitral valve injury is more likely than in the adult population. However, in our and in the authors' experience, septal myectomy remains safe and effective in symptomatic children and late survival is better than the previously published untreated natural history of HCM (2, 18, 19). In adults, alcohol septal ablation can be an alternative to surgery, but there is a general consensus that it should not be used in children due to concerns relating pro-arrhythmia and technical limitations. Isolated case reports and a case series published have suggested that an alternative technique, endocardial radiofrequency septal ablation may be useful in the paediatric population, but further studies are necessary (20).

Sudden cardiac death is the most important clinical problem in hypertrophic cardiomyopathy, may be the first manifestation of the disease. The results of recent work has shown that the highest risk of SCD occurs in the period before puberty (9-11.9 yrs = 7.5% per year) and during early adolescence (12-15.9 yrs = 4.6% per year), and in the age of 16-19.9 years is 1.8% per annum (21). It is worth emphasizing that the average age of our children who died suddenly was 11 years (range 8.3 to 13 years), and so was within the age of highest risk of SCD. In the opinion of experts (11-13) in patients with HCM after resuscitated cardiac arrest and in patients at high risk of SCD, in which $\geq$ 2 major risk factors of SCD are present, the most effective treatment is the implantation of ICD. In study group ICD was implanted in 15.7% of children, in 5 (4.6%) as secondary prevention, while 11% in the primary prevention of SCD. Other authors demonstrated that in 10% of children ICD for primary prevention of SCD was implanted, which is comparable with the results of their own (22, 23). Expert reviews are not fully consistent regarding the indications for ICD implantation for primary prevention in patients with one risk factor for SCD. According to the guidelines of the 2006, ICD implantation can be effective as primary prevention

of SCD in patients with HCM who have one or more major risk factors for SCD (12). The ACC/AHA guidelines in 2011 suggested ICD implantation in primary prevention of SCD in patients with at least one major risk factor such as syncope, massive LV hypertrophy ≥ 30 mm and the occurrence of SCD in the family history, but in the case of the presence of such factors as the NSVT or ABPR during exercise there must be in addition other major factors or modifying factors (13). In our group, in 6 patients with one major risk factor ICD in primary prevention of SCD was implanted. It should be stressed that these children had the major risk factors as syncope, sudden cardiac death in the family due to HCM, massive LV hypertrophy ≥ 30 mm, which in the guidelines of 2011 were considered relevant and sufficient to qualify patients with HCM to ICD implantation. Although risk factors for SCD in adult have been well described (11-13), it seems that they suboptimally differentiate children at high and low risk of SCD (24). Therefore, the ICD implantation should be planned with great caution, especially in pediatric population. The literature underlines that the number of heart transplants performed in children with HCM and severe heart failure is still very low and ranges from 1% to 4% of patients (2, 4, 23, 25). In the study group in a mean follow-up period of 8.2 years died 11 (10.6%) patients. The average annual mortality rate was 1.34, which is comparable with the results of other authors (2, 22).

CONCLUSIONS

In most children with HCM lifestyle modification and appropriate pharmacological therapy are sufficient, additional medical interventions are not necessary. In patients with symptomatic LVOTO refractory to medical therapy a surgical myectomy should be considered. High-risk patients with HCM ought to be prospectively identified and ICD implantation should be considered, however further research is required to establish better criteria for primary prevention in children.

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