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SUPRAVENTRICULAR CARDIAC ARRHYTHMIAS AND SICK SINUS SYNDROME IN PATIENTS WITH A UNIVENTRICULAR HEART AFTER FONTAN PROCEDURE FOLLOW-UP STUDY

This paper describes supraventricular cardiac arrhythmias and sick sinus syndrome in patients with univentricular heart defects, following Fontan procedure. The Fontan operation is a well established surgical procedure in which the right atrium is connected with the pulmonary artery. This technique relieves the functional problems resulting from having a univentricular heart and it is also associated with a lot of complications. Results from 29 patients were included in our study for analysis. All patients with single ventricle were diagnosed and hospitalized in the Department of Paediatric Cardiology in Katowice during 1994–2008. Supraventricular cardiac arrhyhtmias and sick sinus syndrome were common complications in children who went through Fontan procedure. All the stages of treatment with the Fontan procedure are palliative in their character and their main advantage is the improvement in the quality and length of life.

1. BACKGROUND

Many congenital heart defects, in which a heart consists of only one fully developed ventricle, are described as a univentricular heart. The term "functionally single ventricle" is generally used in the clinical practice. The most often observed types of the univentricular hearts are: double inlet left ventricle (DILV), tricuspid atresia (TA), double inlet right ventricle (DIRV), hypoplastic left heart syndrome (HLHS) and the undifferentiated form of the complete atrioventricular canal [9,13] The univentricular heart occurs in approximately 8–10% of children with congenital heart defects [13]. In terms of structure the single ventricle is divided into morphologically left systemic ventricle (70–75%), morphologically right systemic ventricle (25–30%) and into morphologically undifferentiated. Characteristic feature of a univentricular heart is the coexistence of the circulatory system defects [13].

In each anatomical type of the defect the clinical symptoms depend on the size of the pulmonary flow which determines the first stage of cardiosurgical treatment (the banding of the pulmonary artery or systemo–pulmonary anastomosis). Natural history shows that 70% of children die at the age of one year. The procedure performed by Fontan and Baudet in 1971 became the crucial event in the treatment of an univentricular heart [10,11]. Since 1971, numerous modifications of this method have been used in the therapy of congenital heart defects in which biventricular correction is impossible. The Fontan operation is a palliative procedure which connects the right atrium with the pulmonary artery for gas exchange because the systemic and pulmonary circulations are completely separated. The aim of the Fontan procedure is to achieve adequate blood oxygen saturation and to relieve the univentricular heart [10]. Until 1989, the procedure was performed only in one stage. The return of blood from the systemic veins was directed to the pulmonary circulation. Due to many complications associated with this technique, it was divided into two stages (first stage – Hemi–Fontan operation – the connection of the vena cava superior with pulmonary artery and second stage – the Fontan procedure – putting in the drainage of vena cava inferior to the pulmonary circulation) [11,13]. This technique, regardless of its unquestionable advantages still is associated with a lot of complications such as transudates to the serous cavities, thrombo–embolic incidents, dysfunction of heart rhythm and progressive ventricular dysfunction.

The main cause of the cardiac arrhythmias is an organic arhythmic ground (elevated central venous pressure, the lines of the cut in the right atrium and the anastomoses in the vicinity of the sinus node, not sufficient tissue protection, extracorporeal circulation) the other causes are hemodynamics instability, hypoxia, the lack of acid-base balance, water balance, electrolyte balance and drugs, especially catecholamines. Most often observed are arrhythmias due to disorder of the sinus node function, atrial tachycardia and atrial flutter. These crucial cardiac arrhythmias may compound insufficiency of circulation and lower tolerance to exertion and lead to severe cardiac insufficiency and death. That is why cardiac arrhythmias are important clinically and patients suspected of cardiac arrhythmias require cardiological control. [4,5,6].

2. INVESTIGATION PURPOSE

The aim of our study was to conduct a retrospective analysis of the supraventricular heart rhythm disturbances observed in children with an univentricular heart, after completion of cardiosurgical treatment (Fontan procedure).

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3. MATERIALS AND METHODS

The Fontan procedure was performed in 29 children: 11 girls (38%) and 18 boys (62%) between the ages of 1,5 and 17 years (mean age was 8) hospitalized in The Clinic of Children Cardiology between 1994–2008. These cardiosurgical procedures were performed in Katowice during 1994–2007. The follow-up time period was 1 month–14 years. Twenty one percent (21%) of children went through a single-stage repair and 79% of them went through a double-stage repair procedure. In our study, we observed the following types of malformation in children: 44,8 % presented with tricuspid atresia (TA), 34,5% presented with double inlet left ventricle (DILV), 10,3% presented with hypoplastic left heart syndrome (HLHS) and 10,3% presented with different malformation connected with single ventricle.

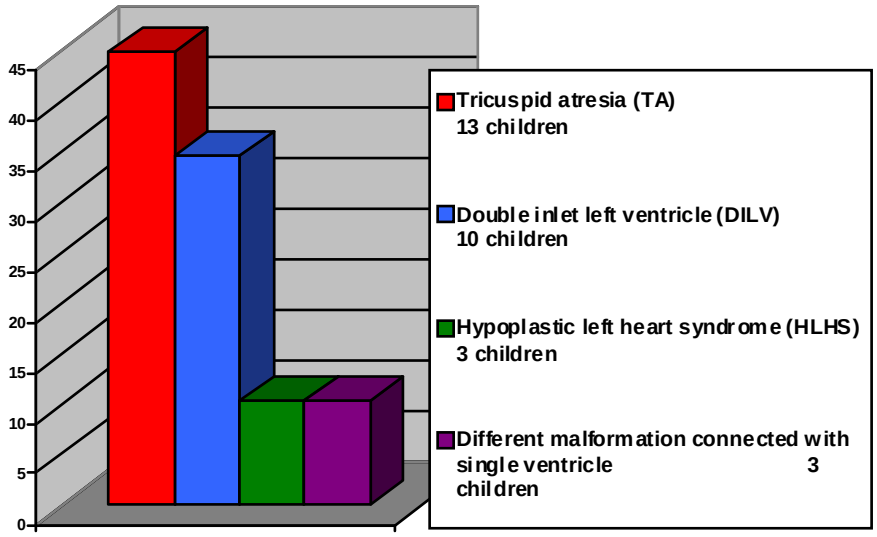

Fig.1 Types of structural heart defects in study group

The results of routine ECG, 24 hour ECG, echocardiography, laboratory findings and imaging studies of the cardiac stress test were analysed.

4. OBTAINED RESULTS

On the basis of ECG and 24 hour ECG results three subgroups were distinct based on presenting symptoms.

I – symptomatic heart rhythm dysfunction observed in 11 patients (38%)
II – asymptomatic heart rhythm disturbance observed in 12 patients (42%)
III – without heart rhythm disturbance observed in 6 patients (20%)

Table 1 lists the types of heart rhythm disturbances, observed in the children from groups I and II.

Table 1. Types of arrhythmias in group I and group II

Type of arrhythmia	group I (*n*)	group II (*n*)
Sick sinus syndrome (SSS)	8	–
Paroxysmal atrial tachyarrhythmia	3	–
Sinus bradycardia, escape rhythms	10	12
Premature supraventricular extrasystolia	4	6

According to Garson criteria for diagnosis of sick sinus syndrome is finding in the electrocardiogram: relevant sinus arrhythmia, sinus bradycardia, sinus arrest, slow escape rhythm, sinoatrial block I, II type 1 and 2, III or tachy-brady syndrome. [4]In the clinical practice not every patient suffering from sinus bradycardia is diagnosed as sick sinus syndrome. Some cases of benign forms of sinus bradycardia require further clinical observation and are not defined as sick sinus syndrome.Other heart rhythm disturbances, seen in the ECGs of children from group I and II are described below.

In group I, five (5) patients had symptomatic arrhythmias (low tolerance of physical exertion, increase in cardiac insufficiency). Six (6) children (3 with sick sinus syndrome and 3 with paroxysmal atrial tachyarrhythmias) required cardiac pacing. Three (3) children had cardioversion and one (1) patient required catheter ablation. Four (4) children were treated chronically with antiarrhythmics.

The heart rhythm disturbances of children from group II were asymptomatic and did not require treatment. Table 2 lists frequency of individual heart rhythm disturbances in children after Fontan operation was performed.

Table 2. Frequency of arrhythmias related to the type of Fontan operation

Type of Fontan operation	Group I, n=11 Significant cardiac arrhythmias	Group II, n=12 Non significant cardiac arrhythmias	Group III, n=6 Without cardiac arrhythmias
Single-stage repair	*n*=3, (27%)	*n*=3, (25%)	–
Double-stage repair	*n*=8 ,(73%)	*n*=9, (75%)	*n*=6, (100%)

* Notice: Fisher's test calculations are attached in the appendix

Heart rhythm disturbances occurred in each child operated on with the classical procedure (one stage). Three (3) children from group I had relevant arrhythmias and 3 children from group II had non relevant arrhythmias. Seventeen (17) patients who received the double stage repair procedure (74%) presented with heart rhythm disturbances but only 8 of them (34%) had significant arrhythmias.

Each patient, with morphologically right ventricle (6 children) had arrhythmias, and 4 of them are included to group I. Four (4) children from group I and II presented with arrhythmias during the first year after operation. In group of 6 children arrhythmias occurred from the first year to the third year after operation. In group of 13 children cardiac arrhythmias occurred after 3 years after operation and were progressively increasing.

5. DISCUSSION

Before the introduction of the condition of the patients was progressively aggravating in terms of management, hypoxia and dysfunction of the ventricle rapidly increased, which leaded to the immediate death of the patient during early childhood.

The aim of Fontan procedure is to improve the quality of life and to extend its length. The procedural method utilized by the Fontan and Baudet operation was to connect the right atrium with the pulmonary artery. This well established surgical procedure was the initial important event in the treatment of the patients with single ventricle. It provided improvement of the clinical condition in patients with an univentricular heart, decrease the level of cyanosis and improved overloading of the systemic ventricle [11,13]. Consequences associated with the Fontan procedure were increasing central venous pressure, overload of the right atrium and its dilatation, which could causes the supraventricular cardiac arrhythmias [6,7]. The Fontan operative techniques have progressively been modified, to minimize these consequences.

One modification, the double-stage intracardiac de Leval's technique (TCPC–total cavopulmonary connection) restricted the area of the increased central venous pressure, reduced progressively volume of the ventricle and leaded to decrease in the postoperative complications [10]. The difference previously observed between the classical stage 1 procedure and double stage techniques, were also observed in our study population. Six (6) patients who went through the classical one–stage repair, suffered from arrhythmia. In the group of 23 children operated in two stages, 17 patients (74%) presented with arrhythmias (crucial heart rhythm disturbances occurred in 8 patients (34%). According to Gellat, 5 years after one-stage repair, crucial heart rhythm disturbances occurred in 30% of the patients. Five (5) years after intracardiac total cavopulmonary connection, arrhythmia was present in 14% of patients [1].

Sick sinus syndrome (8 patients) was observed most often in our patient population. In papers [5,6] attribute sick sinus syndrome observed in patients to the vessels' cannulation and scars after anastomoses in the right atrium. Finally, the scars in the area of the right atrium and its dilatation may lead to atrial tachyarrhythmias which can cause the acute insufficiency of the heart and sudden death. The treatment of choice is cardioversion [12]. Ablation is also commonly used method of treatment [4,12].

One of the most modern modifications to the Fontan procedure is extracardiac total cavopulmonary connection which enables the most laminar flow of blood from the venous system to the pulmonary artery [3,5]. Reduction of blood pressure in the systemic atrium, decrease of the atrial wall's tension and lack of the stitch in its wall may lead to the decrease in the incidence of postoperative arrhythmias which is clinically proved [3,5].

However, in our group of patients total extracardiac cavopulmonary connection was not performed. Therefore, we were not able to observe the benefits of this modification.

Many authors [8,9,10] emphasize that the morphological type of the heart defect influences the incidence of postoperative complications. It has been proved that in the course of time the systemic ventricle in the Fontan circulation will become hypertrophic. This process is more progressive in the morphologically right systemic ventricle [8].

In our study each child with the morphologically right systemic ventricle (6 patients) had heart rhythm disturbances. According to the medical literature, the age of the children and the length of observation of the patients after operation influences the frequency of arrhythmias incidence. According to Gentles supraventricular tachyarrhythmias occurred in about 6% of the patients after 2 years after operation, in 12% after 5 years and in 33% after 10 years after operation [2].

In our group 57% of the children presented the heart rhythm disturbances which occurred after the third year after operation and aggravated in the course of observation. Children with Fontan circulation represent a very demanding and difficult group of patients. Some of the problems associated with these patients can be attributed to their age, since these patient present for treatment during the early years of development. Other problems are associated directly with the extensive

diagnostic evaluation, treatment and surgery required by this young population. All patients after Fontan procedures must be in a controlled environment, in a specialized hospitals with significant clinical experience, where imaging methods are available, ablation, electrophysiological and cardiological procedures are performed.

In the course of the improvement of the operative techniques and the progress of knowledge about circulation pathophysiology the amount of complications after each stage of operation has decreased. It is important to emphasize that all the stages of treatment in patients with single ventricle are palliative in their character and the definitive therapy is heart transplantation.

6. CONCLUSIONS

1. Supraventricular heart rhythm disturbances and sick sinus syndrome are common complications in children who went through the Fontan procedure.
2. Arrhythmias occur more often in single-stage operated children, with morphologically right systemic ventricle.
3. All patients who went through the Fontan procedures, requires systematic cardiological control.

APPENDIX

Gathered in the Table 2 investigation results can be verified by means of the Fisher-Snedecor test. This method is also known as the F-test. In practice, this statistics can be also programmed by means of the ANOVA procedure, which is included in the statistical toolbox of the MATLAB package.

For clarity, calculations step by step were performed.

In the first stage data from Table 2 can be re-written as form:

Result of the operation	Fontan operation type I x_1	Fontan operation type II x_2
G I	$x_{11} = 3$	$x_{21} = 8$
G II	$x_{12} = 3$	$x_{22} = 9$
G III	–	x_{23}=6

In the next step the null hypothesis H_0 can be formulated: mean results of the operations are the same. In other words we assume that:

$$H_0 = \mu_1 = \mu_2$$

Additional, we assume, that the groups x_1 and x_2 have normal distribution and their variances are equal: $\delta_1^2 = \delta_2^2$.

Let n_1 and n_2 be a number of elements in the groups x_1, x_2, respectively. Hence, $n_1 = 2$, $n_2 = 3$.

Let k be a number of the groups. For mentioned above assumptions the Fisher's test can be carried out:

The total number of elements in the groups x_1 and x_2: $N = \sum n_i$.

The mean of the samples in the group i: $\overline{x}_i = \dfrac{\sum_{j=1}^{n_i} x_{ij}}{n_i}$. The mean of the all samples in the groups:

$$\overline{x} = \frac{\sum_{j=1}^{n_1} x_{1j} + \sum_{j=1}^{n_2} x_{2j} + \ldots + \sum_{j=1}^{n_p} x_{kj}}{N}$$

The variances inside groups:

$$\delta'^2 = \frac{\sum_{j=1}^{n_1} (x_{1j} - \overline{x}_1)^2 + \ldots + \sum_{j=1}^{n_p} (x_{kj} - \overline{x}_k)^2}{N - k}$$

and the variances between groups:

$$\delta''^2 = \frac{\sum_{j=1}^{k} n_j (\overline{x}_j - \overline{x})^2}{k-1}$$

In our case we have: $n_1 = 5-2 = 3$ and $n_2 = 2-1$. In the simple calculations we obtain results: $\overline{x}_1 = 6/2 = 3$, $\overline{x}_2 = 23/3 = 7.7$, $\overline{x} = 29/5 = 5.8$ and $\delta'^2 = 1.56$, $\delta''^2 = 26.51$

After calculation, *F*-statistics can be calculated:

$F = \frac{\delta''^2}{\delta'^2} \approx 17$. From appropriate table of the *F*-distribution for $n_1 = 2$ and $n_2 = 3$ degrees of freedom and significance level 0,05 critical number for this parameters is $F_{0,05} = 10.13$. Because $F_{0,05} = 10.13 < F = 17$, null hypothesis should be rejected. Hence, results of the operations are not the same and one type operation gives better results than the second. In other words, double-stage repair Fontan operation gives better results and less complications for patients than single-stage Fontan procedure.

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