УДК 616.12-008.318:616.124-092]-053.2
DOI: 10.20538/1682-0363-2018-2-60–70
For citation: Plotnikova I.V., Dzhaffarova O.Yu., Svintsova L.I., Saushkin V.V., Kovalev I.A. Arrhythmias in children with a single ventricle. *Bulletin of Siberian Medicine*. 2018; 17 (2): 60–70.

Arrhythmias in children with a single ventricle

Plotnikova I.V.[1], Dzhaffarova O.Yu.[1], Svintsova L.I.[1], Saushkin V.V.[1], Kovalev I.A.[2]

[1] *Research Institute Cardiology, Tomsk National Research Medical Center (TNRMC) of Russian Academy of Sciences (RAS)*
111a, Kievskaya Str., Tomsk, 634012, Russian Federation

[2] *Research and Clinical Institute for Pediatrics*
2, Taldomskaya Str., Moscow, 125412, Russian Federation

ABSTRACT

The paper presents the findings on the prevalence rate and structure of arrhythmias and conduction disorder in children with chronic heart defect (CHD) and a functionally univentricular heart (FUH) at the stages of hemodynamic correction. The article also analyses factors determining the risk of arrhythmia development. Seventy patients with CHD and FUH who underwent all the stages of hemodynamic correction were examined. At each stage of surgical correction all children had an examination, including cardiac ultrasound, pulse oximetry, cardiac catheterization and angiopneumography, ECG, and 24-hour ECG monitoring before and after surgery, in order to reveal arrhythmia and conduction disorder. Absence of rhythm disturbances at the first stage of hemodynamic correction was 98.1%, at the second – 80%, at the third – 48.6% and at the fourth stage – 41.5%. Starting from the second stage of hemodynamic correction, the most frequent rhythm disturbance was dysfunction of the sinus node. Statistical analysis showed that there was a significant decrease in oxygen saturation in capillary blood (73 $\pm$ 12% and 81 $\pm$ 4%, respectively, $p < 0.05$) and an increase in the average pressure in the pulmonary artery after total cavopulmonary connection (TCPC) (9.6 $\pm$ 4.2% and 11.5 $\pm$ 5.8%, respectively, $p < 0.05$) in the group of children with heart rhythm disturbances as compared to children without arrhythmias. The average age of patients with arrhythmias in the group of patients having TCPC and, respectively, after the closure of fenestration, was statistically significantly higher than in the group of children without arrhythmias (72 $\pm$ 43 months and 52 $\pm$ 27 months, respectively, $p < 0.05$). The study of arrhythmias in patients with a functionally univentricular heart requires prospective observation, since they significantly affect the prognosis of the disease and the quality of life.

Key words: postoperative arrhythmias, children, univentricular heart, stages of hemodynamic correction of congenital heart defect.

Postoperative arrhythmias are a frequent and serious complication after a cardiovascular surgery. Among the conditions associated with the development of life-threatening arrhythmias, the dominant position belongs to congenital heart defects (CHD), including corrected ones. Incidence of different types of arrhythmias, depending on the initial hemodynamic state and the type of CHD correction, ranges from 10% to 60% [1, 2]. Postoperative rhythm disturbances can occur both in the early postoperative period – immediately after the operation or within 30 days after it, and in the distant one – months and years after the CHD correction. Recently, the number of operations

✉ *Dzhaffarova Olga Yu.,* e-mail: oyd@cardio-tomsk.ru.

in patients with a functionally univentricular heart (FUVH) has increased, and its prevalence constitutes about 5% among all cases of CHD [3]. With FUVH, it is impossible to restore the function of a two-ventricular heart, that is why all patients undergo stage surgical treatment aimed to divide the circulation [4,5]. At any stage of surgical treatment, it is possible to develop various complications, including disturbances in heart rhythm and conduction. With the introduction of the modern modification of the Fontan procedure with the use of an extracardiac conduit, the prevalence of arrhythmias in this group of patients has significantly decreased [6]. However, it continues to be one of the most serious complications affecting the functional state of the patient. There are some studies which considered the risk factors for early and late arrhythmias depending on the modification of the Fontan procedure [6, 7]. J.H. Nurnberg et al. revealed that one of the reasons for late arrhythmias is the absence of a sinus rhythm when discharging from a hospital [6]. There are no papers devoted to the analysis of arrhythmia development in one cohort of patients at the stages of hemodynamic correction in case of CHD and FUVH, but it is essential for finding early markers of its development.

The aim of the study is to evaluate the incidence and structure of cardiac arrhythmias and conduction in children with CHD and FUVH at the stages of hemodynamic correction and specify the factors determining the risk of arrhythmias.

PATIENTS AND RESEARCH METHODS

We examined seventy patients with CHD and FUVH, who underwent total cava-pulmonary anastomosis with an extracardiac conduit. The structure of the CHD among the examined patients is presented in Tab. 1.

Table 1

SThe structure and incidence of CHD, including FUVH, among examined patients

Structure	Number absolut (relative, %)
Double-inlet left ventricle	17 (24,3)
Tricuspid atresia	16 (22,9)
Mitral atresia	9 (12,9)
Unbalanced atrioventricular canal defect	7 (10)
Double outlet right ventricle	6 (8,6)
Hypoplastic left-heart syndrome	4 (5,7)

End of table 7

Structure	Number absolut (relative, %)
Pulmonary atresia with intact ventricular septum	2 (2,8)
Ebstein's disease	2 (2,8)
Other FUVHs modifications	7 (10)

Note: *n* – number of patients.

ThAmong patients with FUVH, five (7.14%) children were diagnosed with heterotaxy syndrome, which was combined with left isomerism in four patients and with right isomerism in one patient.

The first stage of hemodynamic correction was performed in 52 (74.3%) children, 22 of them (42.3%) underwent systemic-pulmonary anastomosis (modified Blalock-Taussing shunt), 26 patients (50%) underwent pulmonary artery banding and four (7.4%) children had a Norwood surgery. The remaining 18 patients did not undergo the first stage due to a balanced blood circulation.

At the second stage of hemodynamic correction 68 (97.1%) patients underwent bidirectional cava-pulmonary anastomosis (BCPA), two (2.9%) patients had a Kawashima procedure at the age of 1 and 2 years old respectively.

The third stage of hemodynamic correction, which is the Fontan procedure (total cava-pulmonary anastomosis - TCPA) was performed on all patients. 68 patients (97.1%) underwent a Fontan procedure with an extracardiac conduit, two patients had a transfer of hepatic veins to the pulmonary artery by a "Gore – tex" extracardiac conduit. It should be noted that patients with the Fontan procedure at this stage experienced direct fenestration between the extracardiac conduit and the free wall of the right atrium. Six to nine months after TCPA 65 (92%) patients had the fenestration closed. Two patients did not have the fenestration closed within the control period due to increased pressure in the pulmonary artery, three patients died in the period preceding the implementation of this stage of treatment.

12-24 months and 25-30 months after TCPA, 31 (44.3%) and 33 (47.1%) patients were examined at the point of catamnesis respectively.

It is important to mention that not all patients had surgical interventions performed in the same clinic. The following operations were performed at each stage in the Scientif-

ic Research Institute of Cardiology: stage I - 24 (34.3%), stage II - 50 (71.4%) stage III- 70 (100%); stage IV -65 (92%).

METHODS OF ARRHYTHMIA DETECTION

At each stage, all children underwent a medical examination, which included clinical examination, heart ultrasound, pulse oximetry, cardiac catheterization and angiopulmonography,and an ECG with 12-standard leads before and after surgery. After each operation all patients had bedside ECG monitoring for several days with the aim to detect cardiac rhythm disturbances in a timely manner. ECG daily monitoring of the (ECGDM) was performed before the operations and 5-8 days after them. After all stages of hemodynamic defect correction, all children were recommended to have ECGDM once a year, and if there are any deviations of heart rhythm or conduction, then they would have to have this procedure more often.

Transesophageal electrophysiological examination was performed in case of complaints of palpitations, and also for the evaluation of sinus node function and diagnosis of tachycardia paroxysms. If surgical treatment was performed at certain stages in other clinics, data from medical history extracts and archival ECG and ECGDM data were used to assess arrhythmias. To determine exercise tolerance in order to clarify the functional class (FC), patients who had experienced the third stage of hemodynamic correction underwent a modified Harvard step test [8].

In order to systematize the results of the study, all patients were divided into eight groups:

1 – sinus rhythm (heart rate corresponds to the age norm) [9,10];

2 –non-sinus rhythm (atrial rhythm, nodal rhythm, AV-dissociation with the average daily heart rate within the age limit);

3 – bradyarrhythmia (sinus bradycardia and atrial rhythm with an average daily HR <5 ‰ of the distribution scale for a given age);

4 – sinus tachycardia;

5 – supraventricular tachycardia (paroxysmal and permanent forms);

6 – extrasystole (supraventricular and ventricular)

7 – atrioventricular blockade (AVB) of I-II degree (mean daily heart rate according to SMECG data corresponds to the age norm);

8 – AB blockade of the III degree

STATISTICAL ANALYSIS

Statistical processing of the results was carried out using STATISTICA 10.0 for Windows. The normality of distribution laws for numerical values was verified using the Kolmogorov-Smirnov test. In case of the normal distribution of characteristics, the data were presented in the form (M ± SD), where M is a mean value, and SD is the standard square deviation. The absolute number and the relative value in percent form were indicated for the values characterizing the qualitative characteristics. To assess the reliability of the differences in independent samples, a nonparametric Mann-Whitney U Test was used. To assess the reliability of the differences in dependent samples, a non-parametric Wilcoxon Test was used. The reliability of differences in qualitative characteristics was determined using the Chi-square test. Differences were considered statistically significant at $p < 0.05$.

RESULTS AND DISCUSSION

Patients

Biometric and preoperative ultrasound parameters and catheterization data at the stages of hemodynamic correction of CHD are presented in Table 2

Table 2.

Biometric and preoperative ultrasound parameters and catheterization data at the stages of BCPA, TCPA and fenestration closure

Parameters	BCPA $n = 70$	TCPA $n = 70$	Fenestration closure $n = 65$
Gender	$b = 37\ /g = 33$	$b = 37/g = 33$	$b = 35/g = 30$
Age (months)	15,5 ± 19,49	47,17 ± 17,95	72,87 ± 42,11
Weight (kg)	8,63 ± 3,79	17,13 ± 6,2	19,1 ± 7,15
SpO_2	75,04 ± 9,03	79,51 ± 5,87	84,45 ± 5,24

End of table 2

Parameters	BCPA $n = 70$	TCPA $n = 70$	Fenestration closure $n = 65$
mPAP(mm Hg)	14,16 ± 8,49	9,81 ± 4,35	8,71 ± 2,65
End-diastolic pressure FUVH (mm Hg)	6,62 ±3 ,45	7,81 ± 2,34	6,42 ± 2,09
EF FUVH	67,48 ± 8,07	64,83 ± 7,59	61,33 ± 6,91
Rp	1,26 ± 0,59	1,07 ± 0,66	1,09 ± 0,53
Rs	9,88 ± 4,59	13,04 ± 6,44	12,12 ± 4,22

n – number of patients; *b* – boys; g- girls; Rp- pulmonary vascular resistance; Rs – systemic vascular resistance; SpO_2 – oxygen saturation in the blood; EF – ejection fraction

Among the examined patients there were three (4.3%) lethal cases after the Fontan operation, one of which was early (within 30 days after the operation because of acute cardiopulmonary insufficiency) and two late ones due to progressive venous thrombosis (within 3.5 and 6 months after surgery).

At I stage, only one patient (1.4%) in the early postoperative period (on the 7th and 15th day) had two paroxysms of supraventricular tachycardia, corrected by a bolus injection of cordarone.

At II stage of hemodynamic defect correction (BCPA) we recorded heart rhythm and conduction disturbances in 15 (20%) patients, seven of which (10%) experienced them on admission to the clinic before BCPA (Table 3).

After BCPA, newly observed rhythm and conduction disturbances were registered in eight patients (8.6) %, six patients (8.6%) preserved them, and 56 patients (80) % didn't have any.

Table 3

Structure and incidence of rhythm disturbances and cardiac conduction at the stages of hemodynamic management of children with FUVH

Arrhythmia structure	I stage, *n* (%)	II stage		III stage		IV stage (fenestration closure)	
		before, *n* (%)	after, *n* (%)	before, *n* (%)	after, *n* (%)	before, *n* (%)	after, *n* (%)
Sinus rhythm	51 (98,1)	63 (90)	56 (80)	57 (81,4)	34 (48,6)	25 (38)	27 (41,5)
Non-sinus rhythm	–	5 (7,2)	6 (8,6)	6 (8,6)	9 (12,8)	8 (12)	9 (13,8)
Bradyarrhythmia	–	–	2 (2,8)	–	9 (12,8)	11 (16,9)	11 (16,9)
Sinus node tachycardia	–	–	2 (2,8)	–	5 (7,2)	10 (15,3)	6 (9,2)
SVT	1 (1,9)	1 (1,4)	–	–	6 (8,7)	3 (4,6)	2 (3,0)
Ectopic heartbeat	–	–	1 (1,4)	2 (2,8)	2 (2,8)	3 (4,6)	3 (4,6)
Atrioventricular block I–II st.	–	–	–	2 (2,8)	2 (2,8)	1 (1,5)	3 (4,6)
Atrioventricular block III st.	–	1 (1,4)	3 (4,4)	3 (4,4)	3 (4,4)	4 (6,10)	4 (6,1)
Total number of patients	52	70	70	70	70	65	65

Note: *n* – number of patients, SVT – supraventricular tachycardia.

At III stage of hemodynamic correction (TCPA) in children with FUVH, cardiac rhythm and conduction disturbances were registered in 36 (51.4%) patients receiving the operation, which was statistically significantly higher than at the previous stage ($\chi 2 = 13.6$, $p < 0,05$). Before TCPA procedure, the structure of arrhythmias was practically the same as after the 2nd stage of hemodynamic correction (Table 3).

The structure of arrhythmias after TCPA procedure is presented in Table 3. Sinus tachycardia diagnosed in this group of patients was seen as the activation of the sympathetic part of the autonomic nervous system related to heart failure in the postoperative period. During the treatment we noticed a heart rate fall. Only two patients needed an additional prescription of anaprilin to control their heart rate. At this stage of hemodynamic correction bradyarrhythmias were diagnosed only after TCPA. Four out of nine patients in this group were diagnosed with a sinus bradycardia, the rest of

them had bradyarrhythmia represented by the atrial rhythm with an average daily heart rate below the age norm. In the group of patients with non-sinus rhythm five patients experienced these changes earlier, and four of them had an atrial rhythm due to isomerism. The remaining four patients were diagnosed with atrial rhythm after the surgery. Two patients preserved premature ventricular contraction, detected prior TCPA. One of them had a decrease in ectopic activity of ventricular premature beats (VPB) (from 2% to 0.5%), and the other had the same supraventricular extrasystole (SVES) activity - 16%. Two other patients preserved AVB, and one of them showed AVB progression from I stage to II stage (Mobitz 2). Two patients with AVB of the third degree underwent electric cardiac pacemaker implantation.

Among our patients there were children over the age of 4 who underwent a Fontan procedure (20 patients - 28.6%), and the oldest was 18 years old. We did not find a sinus rhythm in most of these patients (15 out of 20) after TCPA. The absence of a sinus rhythm could be explained by the anatomic features of cardiac conduction system (isomerism) in only 4 cases. Two out of five patients with sinus rhythm had a heart rate that corresponded to the age norms, whereas the other three were diagnosed with bradycardia. Twelve months after TCPA, one patient from this group, who had not been previously diagnosed with heart rhythm disturbances, was diagnosed with an atrial bradycardia.

Thus, after TCPA procedure, new disturbances of heart rhythm and conduction were recorded in 23 (32.9%) patients, 13 patients (18.5%) preserved them, and these disturbances were not registered in 34 patients (48.6%).

At the 4th stage of hemodynamic correction heart rhythms and conduction disturbances were found in 40 (61%) and 38 (58%) patients respectively before and after the fenestration closure. Their structure is shown in Table 3. Before closing the fenestration, heart rhythm and conduction disturbances were almost comparable to the similar disturbances in incidence revealed after carrying out the III stage (TCPA) (Table 3). What is interesting is that in the majority of children with bradyarrhythmias (9 out of 11) bradyarrhythmia emerged in the first days after TCPA and was presented by a delayed atrial or nodal rhythm. Over time one child out of nine was registered with bradycardia progression that manifested in an increasing rhythm pause for more than 2.5 seconds, symptomaticity, weakness episodes, and fatigue that required pacemaker implantation, together with the fenestration closure. All patients with atrial tachycardia were prescribed medication before the fenestration closure: two patients were administered anaprilin, one patient was given digoxin. As a result of treatment, within 2–3 weeks, the average daily heart rate dropped to normal ranges.

After the fenestration closure, the structure of rhythm and conduction disturbances did not change significantly (Table 3). The mean daily heart rate of 2/3 of the children who had sinus tachycardia before the fenestration closure continued to exceed the age indices. One child after the fenestration closure had a sinus bradycardia, and two children initially having bradycardia were registered with a sinus rhythm with a normal heart rate. Two children were added to the group of patients with AVB I-II stages.

12–24 months after TCPA, 31 (42.3%) patients were examined over time. The structure and frequency of cardiac arrhythmias and conduction disturbances were presented in Table 4. Arrhythmias were recorded in 17 (54.8%) patients. Among patients with sinus tachycardia, which was detected in 5 children, anaprilin was needed only in two cases. Following the treatment there was a decrease in the cardiac rhythm to normal numbers. A new case of heart rhythm disturbances (atrial rhythm) was detected only in one patient who had sinus rhythm with an adequate heart rate at all stages of hemodynamic correction. In the remaining 16 patients, observed heart rhythms disturbances were registered earlier.

Table 4

Structure and incidence of rhythm disturbances and cardiac conduction in children with FUVH after hemodynamic management over time

Arrhythmia structure	12–24 months, *n* (%)	25–30 months, *n* (%)
Sinus rhythm	14 (45,3)	17 (51,5)
Non-sinus rhythm	5 (16,1)	1(3,0)
Bradyarrhythmia	4 (12,9)	8 (24,3)
Sinus node tachycardia	5 (16,1)	4 (12,2)
SVT	1 (3,2)	1 (3,0)
Complete atrioventricular block	1 (3,2)	–
Ectopic heartbeat	1 (3,2)	1 (3,0)
Atrioventricular block 1–2 stage		1 (3,0)
Total number of patients	31	33

25–30 months later, 33 (47.1%) patients were examined over time (Table 4). When comparing data on incidence of the analyzed indicators with the previous stage of the survey, changes were noted mainly in groups of children with non-sinus rhythm and bradyarrhythmias. The number of patients with atrial rhythm decreased from five to one due to the emergence of bradyarrhythmia.

In order to find the reasons most likely associated with the development of arrhythmias at the stages of hemodynamic correction of CHD with FUVH, an additional analysis was performed. As probable factors we considered age at the time of BCPA and TCPA procedures, mean pressure in the pulmonary artery (LA), oxygen saturation in the capillary blood, FUVH end-diastolic pressure (EDP), FUVH contractility, resistance of pulmonary and systemic circulation, the morphology of the systemic ventricle (right or left), arrhythmias at the discharge from the hospital and the patient's FC. It should be noted that such indicators as mean pressure in LA, FUVH EDP, resistance of pulmonary and systemic circulation were measured on the basis of catheterization data before surgery. Oxygen saturation in the capillary blood and FUVH contractile ability was assessed 5-7 days before and after the operation.

We found that a statistically significant decrease in oxygen saturation in capillary blood (73 ± 12% and 81 ± 4%, respectively, $p < 0.05$) was noted in the group of children with heart rhythm disturbances in comparison with children without arrhythmia, and an increase in mean pressure in the LA after TCPA (9.6 ± 4.2% and 11.5 ± 5.8%, respectively, $p < 0.05$). After the fenestration closure, these changes in the analyzed groups were leveled. We can assume that one of the causes of arrhythmia after the Fontan procedure is hypoxia, which is considered as one of the pathogenetic mechanisms in the development and progression of arrhythmias and hemodynamic disorders in children with FUVH. Attention is drawn to the fact that the average age of patients with arrhythmias in the group of patients after TCPA and, respectively, after the fenestration closure, was statistically significantly higher than in the group of children without arrhythmias (72 ± 43 months and 52 ± 27 months, respectively, $p < 0.05$).

DISCUSSION

Recently, the number of operations in patients with FUVH has increased. Rhythm disturbances are one of the main risk factors for the development of morbidity and functional disorders after Fontan procedure.

The absence of sinus rhythm in patients with FUVH, loss of atrioventricular synchronicity leads to a decrease in cardiac output, and as a consequence worsens the prognosis in the long term.

According to various sources, the prevalence of atrial arrhythmias after Fontan procedures varies from 10% to 40%. In the structure of rhythm disturbances, dysfunctions of the sinus node associated with bradyarrhythmias, as well as supraventricular tachycardia (nodal ectopic, atrial fibrillation (fibrillation ectopic tachycardia), intracardiac reentry tachycardia) predominate. [11,12,13,14].

Among our patients, after the Fontan procedure, the structure of heart rhythm and conduction disturbances corresponded to the data of foreign colleagues, with the exception of the absence of atrial fibrillation (AF) and paroxysmal reentry of tachycardia in the early postoperative period. We found an atrial fibrillation episode in the patient at the endovascular fenestration closure, which was successfully stopped after the administration of digoxin. When analyzing the incidence of rhythm disturbances and conduction in our study, patients with bradyarrhythmia and non-sinus rhythm predominated, including atrial, nodal rhythm, and AV dissociation. We would like to note that in four patients with atrial rhythm along with FUVH, heterotaxis syndrome with left isomerism was diagnosed, in which the absence of sinus rhythm is an anatomical feature of the structure of the conduction system of the heart.

Many authors believe that the cause of sinus node dysfunction is its immediate intraoperative trauma during the creation of an anastomosis or a violation of its blood supply [11,13]. Sinus node dysfunction may be a trigger mechanism in the development of atrial tachycardia and predisposing factor in the development of atrial flutter [11,13]. In a number of patients with progressive sinus node dysfunction, an implantation of ECP is required. In the case of ECP implantation, DDD regimen is preferred, which improves pulmonary venous return and cardiac output [15]. Among our patients, ECP

implantation in DDD mode in connection with the progression of the sinus node dysfunction was needed for two patients - one at the TCPA stage and one at the fenestration closure stage.

Despite the fact that heart rhythm and conduction disturbances occur quite often after a Fontan procedure, according to a number of authors, absence of bradyarrhythmias after Fontan procedures is found in 83% and 73% for 5 years and 10 years of observation respectively [16].

In addition to bradyarrhythmias, atrial tachyarrhythmias are one of the problems that can lead to a worsening of hemodynamics and increase the risk of sudden death in children with CHD, and in particular with FUVH [17]. In addition, atrial tachycardia in the form of atrial flutter increases the risk of thrombosis and sudden death and worsen hemodynamics in patients with Fontan hemodynamics [18].

In our study there were patients with atrial tachycardias, which occurred at all stages of hemodynamic correction of the FUVH. In most of these children, the average heart rate according to the ECGDM data slightly exceeded the age indices (no more than 20% of the age norm) and did not require medication. According to various studies, postoperative SVT is most often registered in patients with extracardiac Fontan with heterotaxis syndrome and systemic ventricular dysfunction [19]. Among our patients with heterotoxicity, atrial tachycardia with an insignificant increase in mean daily heart rate (no more than 20% in relation to age standards) was registered in 2 out of 5 patients after TCPA application.

According to a number of authors, after Fontan operation, SVTs were recorded in 5.5% of cases [15]. The complete absence of SVT, in comparison with bradyarrhythmias, was greater and in 5 years was 94% and 90% after 10 years of follow-up [16]. The development of atrial tachycardia is promoted by progressive stretching of the atrial wall, as well as the presence of intra-atrial suture lines [12,14], which is more relevant to patients with intracardial Fontan [12,20].

Most centers report a lower level of occurrence of postoperative atrial tachycardias in patients with an extracardiac conduit, but this complication cannot be completely avoided at this stage, as evidenced by the data of our clinic. The incidence of rhythm disturbances and conduction of the heart after the operation of the extracardiac Fontan was 51.4%. It should be noted that in dynamics at 12 and 30 months arrhythmias were present in those patients in whom these disorders were detected after superimposing TBCs. Our data are not yet numerous due to the fact that not all patients reached the analyzed periods of observation, but, nevertheless, we can correlate them with the results of a number of authors. In particular, in the work of our colleagues who analyzed the risk factors for early SVT after Fontan operation, the presence of arrhythmia at discharge from the hospital contributes to the occurrence of arrhythmias in the future [6].

Comparative data on the frequency of arrhythmia after the extracardiac and lateral Fontan are very contradictory. Colleagues from the Slovak clinic conducted a retrospective analysis of the occurrence of arrhythmias during the year after Fontan operation using various modifications (lateral tonal, or extracardiac conduit). In the course of the study it was found that the number of arrhythmias occurring in the early postoperative period did not differ significantly, and the most frequent violation of rhythm activity was the nodal rhythm [7]. On the contrary, the data presented by the clinic of the Berlin Cardiology Center showed that in the early postoperative period, the percentage of sinus rhythm preservation was 86% and 50%, respectively, in patients with extracardiac Fontan compared with patients who underwent intra-atrial Fontan (p <0.001). There was also a significant reduction in the number of patients with sinus node dysfunction, and as a consequence, the need for implantation of ECS in the group with extracardiac Fontan [8].

M. Dilawar et al. [21] demonstrated that dysfunction of the sinus node is much more common in patients after extracardiac Fontan (59%), whereas in patients with atrial atone it was detected only in 21% of cases. This fact was explained by a longer (twice) observation of patients with extracardiac Fontan. According to the results of one large study [22], where an analysis of 520 patients performed different Fontan modifications, it was shown that the occurrence of arrhythmias was detected in 19% of patients with lateral Fontan and only 2% with an extracardiac conduit . The authors note that the occurrence of rhythm disturbances increases with the age of the patient and further per-

spective observation is necessary to evaluate the long-term results. P.A. Anderson et al. argues that in the case of a Fontan operation before the age of 3 years, the sinus rhythm persists in 70-74% of cases, which was statistically significantly higher than in older patients (62%). These data were presented after analysis of outcomes of surgical treatment in 546 patients from various US clinics [23].

Our data correspond to these studies. In the group of patients with arrhythmias, the age of TPSS was significantly higher than in patients without heart rhythm disturbances. ($p < 0.05$).

In the treatment of arrhythmias in patients after Fontan operation, various methods can be used: drug therapy, radiofrequency ablation, implantation of antiarrhythmic devices [24,25,26]. Antiarrhythmic drugs in patients after Fontan should be used with caution.

Possibilities of pharmacological control of atrial reentry of tachycardia are rather limited - the effectiveness of therapy, including partial therapy rarely reaches 70% [27]. Mono or combined antiarrhythmic therapy of atrial reentry tachyarrhythmia includes blockers of Na-channels, □-adrenoblockers, potassium channel blockers, calcium antagonists, digoxin. The use of blockers of Na-channels (I C subgroup) and K-channels (amiodarone, sotalol) was the most widely spread from the point of view of effectiveness and minimal negative effect on the contractile function of the FUVH [27]. In case of impossibility of drug control of tachyarrhythmia or development of side effects from therapy with antiarrhythmic drugs, the performance of catheter radiofrequency ablation of the arrhythmia substrate has been demonstrated, which has proved its effectiveness and safety, including in children up to one year [28,29,30]. In our study there were patients with atrial tachycardias that arose after the Fontan operation, but they did not need RFA. Only two patients were prescribed therapy with a sotalex with a positive effect at the time of discharge from the hospital. In the remaining children, the average heart rate according to the ECGDM data slightly exceeded the age indices. Syndrome of weakness of the sinus node may become an indication for the implantation of the pacemaker. This occurs if there is a "tachi-bradycardia syndrome" and when establishing a connection of symptoms with an inappropriate age bradycardia [30], which is problematic for patients with FUVH after separation of the circling circles because the use of standard approaches for the diagnosis of bradycardia in such patients is obviously not correct.

CONCLUSION

1. Patients with extracardiac Fontan at all stages of hemodynamic correction had irregularities in rhythm and conduction of the heart. Freedom from arrhythmias amounted to 98.1% at the first stage of correction, 80% at the second stage, 48.6% at the third stage, and 41.5% at the fourth stage.

2. Beginning with the second stage of hemodynamic correction in patients with a functionally single ventricle of the heart, the most frequent arrhythmias were dysfunctions of the sinus node in the form of bradyarrhythmias and atrial rhythms.

3. After the Fontan operation in the group of patients with heart rhythm disorders, in contrast to patients without arrhythmias, there was a statistically significant decrease in oxygen saturation in the capillary blood and an increase in mean pulmonary artery pressure, which may be due to hypoxia, which is regarded as one of the pathogenetic mechanisms in the development and progression of arrhythmias and hemodynamic disorders in the functionally single ventricle of the heart.

4. The average age of patients with arrhythmias in the group of patients after Fontan surgery and, respectively, after the closure of fenestration, was statistically significantly higher than in the group of children without arrhythmias, which may be due to the presence of long-lasting bulk heart overload after performing the III stage of hemodynamic correction.

CONFLICT OF INTEREST

The authors declare the absence of obvious and potential conflicts of interest related to the publication of this article.

SOURCE OF FINANCING

The authors state that there is no funding for the study.

CONFORMITY WITH THE PRINCIPLES OF ETHICS

The study approved by the local ethics committee under the Research Institute Cardiology.

Received 22.12.2017
Accepted 24.04.2018

Plotnikova IrinaV., DM, Head of the Department of Pediatric Cardiology, Research Institute Cardiology, TNRMC RAS, Tomsk, Russian Federation.

Dzhaffarova Olga Yu., PhD, Senior Researcher, Department of Pediatric Cardiology, Research Institute Cardiology, TNRMC RAS, Tomsk, Russian Federation.

Svintsova Liliya I., PhD, Leading Researcher, Department of Pediatric Cardiology, Research Institute Cardiology, TNRMC RAS, Tomsk, Russian Federation.

Saushkin Vyacheslav V., PhD, Senior Researcher, Laboratory of Radionuclide Research Methods, Research Institute Cardiology, TNRMC RAS, Tomsk, Russian Federation.

Kovalev Igor A., DM, Professor, Deputy Chief Physician, Research and Clinical Institute for Pediatrics, Moscow, Russian Federation.

(✉) **Dzhaffarova Olga Yu.**, e-mail: oyd@cardio-tomsk.ru.

REFERENCES

1. Rękawek J., Kansy A., Miszak-Knecht M. et al. Risk factors for cardiac arrhythmias in children with congenital heart disease after surgical intervention in the early postoperative period. *The Journal of Thoracic and Cardiovascular Surgery*. 2007; 133 (4): 900–904.
2. Walsh E.P. Interventional electrophysiology in patient with congenital heart disease. *Circulation*. 2007; 115: 3224–3234.
3. Mavroudis C., Backer C.L. Pediatric cardiac surgery. 3td ed. Mosby, 2003: 875.
4. Hancock Friesen C.L., Forbess J.M. Surgical management of the single ventricle. *Progress in Pediatric Cardiology*. 2002; 16: 47–68.
5. Petit C.J. Staged single-ventricle palliation in 2011: outcomes and expectations. *Congenit Heart Dis*. 2011; 6 (5): 406–416.
6. Nurnberg J.H., Ovroutski S., Alexi-Meskishvili V. et al. New onset arrhythmias after the extracardiac conduit Fontan operation compared with the intraatrial lateral tunnel procedure: early and midterm results. *Ann. Thorac. Surg*. 2004; 78: 1979–1988.
7. Hakacova N., Lakomy M., Kovacikova L. et al. Arrhythmias after Fontan operation: comparison of lateral tunnel and extracardiac conduit. *Journal of Electrocardiology*. 2008; 41: 173–177.
8. Тупикина А.А., Плотникова И.В., Ковалев И.А. и др. Определение толерантности к физической нагрузке у здоровых детей с использованием модифицированного Гарвардского степ-теста. *Сибирский медицинский журнал*. 2015; 30 (4): 36–39. [Tupikina A.A., Plotnikova I.V., Kovalev I.A. et al. Determination of exercise tolerance in healthy children using the modified Harvard step test. *Siberian Medical Journal*. 2015; 30 (4): 36–39 (in Russ.)].
9. Макаров Л.М. Холтеровское мониторирование. 3-е изд. М.: Медпрактика, 2008: 456. [Makarov L.M. Holter monitoring. 3rd ed. M.: Medpraktika Publ., 2008: 456 (in Russ.)].
10. Аритмии у детей. Атлас электрокардиограмм; под ред. М.А. Школьниковой. М.: ИД «Медпрактика-М», 2006; 148. [Arrhythmia in children. The atlas of electrocardiograms; ed. M.A. Schoolboy. M.: Publishing house «Medpraktika-M», 2006; 148 (in Russ.)].
11. Cohen M.I., Wernovsky G., Vetter V.L. et al. Sinus node function after a systematically staged Fontan procedure. *Circulation*. 1998; 98(suppl. II): 352–383.
12. Fishberger S.B., Wernovsky G., Gentles T. et al. Factors that influence the development of atrial flutter after the Fontan operation. *J. Thorac. Cardiovasc. Surg*. 1997; 113: 80–86.
13. Manning P.B., Mayer J.E., Wernovsky G. et al. Staged operation to Fontan increases the incidence of sinoatrial node dysfunction. *J. Thorac. Cardiovasc. Surg*. 1996; 111: 833–840.
14. Durongpisikul K., Porter C., Cetta F. et al. Predictors of early- and late-onset supraventricular tachyarrhythmias after Fontan operation. *Circulation*. 1998; 98: 1099–1107.
15. Drago F., Silvetti M.S., Grutter G., De Santis A. Long term management of atrial arrhythmias in young patients with sick sinus syndrome undergoing early operation to correct congenital heart disease. *Europace*. 2006; 8: 488–494.
16. Kaulitz R., Hofbeck M. Current treatment and prognosis in children with functionally univentricular hearts. *Arch. Dis. Child*. 2005; 90: 757–762.
17. Durongpisitkul K., Porter C.J., Cetta F. et al. Predictors of early- and late-onset supraventricular tachyarrhythmias after Fontan operation. *Circulation*. 1998; 98: 1099–1107.
18. Ghai A., Harris L., Harrison D.A. et al. Outcomes of late atrial tachyarrhythmias in adults after the Fontan operation. *J. Am. Coll. Cardiol*. 2001; 37: 585–592.
19. Chowdhury U.K., Airan B., Kothari S.S. et al. Specific issues after extracardiac fontan operation: ventricular function, growth potential, arrhythmia, and thromboembolism. *Ann. Thorac. Surg*. 2005; 80 (2): 665–672.
20. Cecchin F., Johnsrude C.L., Perry J.C. et al. Effect of age and surgical technique on symptomatic arrhythmias after

the Fontan procedur. *Am. J. Cardiol*. 1995; 76: 386–391.
21. Dilawar M., Bradley S.M., Saul J.P. et al. Sinus node dysfunction after intraatrial lateral tunnel and extracardiac conduit Fontan procedures. *Pediatr. Cardiol*. 2003; 24: 284–288.
22. Stephenson E.A., Lu M., Berul C.I. et al. Arrhythmias in a contemporary Fontan cohort: prevalence and clinical associations in a multicenter cross-sectional study. *J. Am. Coll. Cardiol*. 2010; 56: 890–896.
23. Anderson P.A., Sleeper L.A., Mahony L. et al. Contemporary outcomes after the Fontan procedure. *J. Am. Coll. Cardiol*. 2008; 52 (2): 85–98.
24. Guccione P., Paul T., Garson A. et al. Long-term follow up of amiodarone therapy in the young: continued efficacy, unimpaired growth, moderate side effects. *J. Am. Coll. Cardiol*. 1990; 15: 1118–1124.
25. Beuafort-Krol G.C.M., Bink-Boelkens Th.E. Effectiveness of sotalol for atrial flutter in children after surgery for congenital heart disease. *Am. J. Cardiol*. 1997; 79: 92–94.
26. Chetaille P., Walsh E.P., Triedman J.K. Outcomes of radiofrequency catheter ablation of atrioventricular reciprocating tachycardia in patients with congenital heart diseas. *Heart Rhythm*. 2004; 1: 168–173.
27. Fujta S., Takahashi K., Takeuchi D. et al. Management of late atrial tachyarrhythmia long after Fontan operation. *J. of Cardiology*. 2009; 53: 410–416.
28. Ковалев И.А., Марцинкевич Г.И., Попов С.В. и др. Клиническая эффективность радиочастотной абляции и ее влияние на внутрисердечную гемодинамику у детей. *Тихоокеанский медицинский журнал*. 2004; 4: 16–17. [Kovalev I.A., Marcinkevich G.I., Popov S.V. Clinical effectiveness of radiofrequency ablation and its influence on intracardiac hemodynamics in children. *Pacific Medical Journal*. 2004; 4: 16–17 (in Russ.)].
29. Попов С.В., Свинцова Л.И., Ковалев И.А. и др. Эффективность эндокардиальной радиочастотной абляции тахиаритмий у детей первого года жизни. *Вестник аритмологии*. 2012; 67: 5–10. [Popov S.V., Svintsova L.I., Kovalev I.A. Efficiency of endocardial radiofrequency ablation of tachyarrhythmias in children of the first year of life. *Herald of Arrhythmology*. 2012; 67: 5–10 (in Russ.)].
30. Brugada J., Blom N., Sarquella-Brugada G. et al. European Heart Rhythm Association, Association for European Paediatric and Congenital Cardiology: Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group Joint Consensus Statement. *Europace*. 2013; 15 (9): 1337–1382.

УДК 616.12-008.318:616.124-092]-053.2
DOI: 10.20538/1682-0363-2018-2-60–70
Для цитирования: Плотникова И.В., Джаффарова О.Ю., Свинцова Л.И., Саушкин В.В., Ковалев И.А. Аритмии у детей с функционально единственным желудочком сердца. *Бюллетень сибирской медицины*. 2018; 17 (2): 60–70.

Аритмии у детей с функционально единственным желудочком сердца

Плотникова И.В.[1], Джаффарова О.Ю.[1], Свинцова Л.И.[1], Саушкин В.В.[1], Ковалев И.А.[2]

[1] *Научно-исследовательский институт (НИИ) кардиологии, Томский национальный исследовательский медицинский центр (ТНИМЦ) Российской академии наук (РАН)*
Россия, 634012, г. Томск, ул. Киевская, 111а

[2] *Научно-исследовательский клинический институт (НИКИ) педиатрии имени академика Е.Ю. Вельтищева Российского национального исследовательского медицинского университета (РНИМУ) имени Н.И. Пирогова*
Россия, 125412, г. Москва, ул. Талдомская, 2

РЕЗЮМЕ

Цель исследования – изучить распространенность и структуру нарушений ритма и проводимости сердца у детей с врожденным пороком сердца (ВПС) с функционально единственным желудочком сердца (ФЕЖС) на этапах гемодинамической коррекции порока, а также проанализировать факторы, определяющие риск развития аритмий.

Материалы и методы. Обследовано 70 пациентов с ВПС с ФЕЖС, которым были выполнены все этапы гемодинамической коррекции порока. Для выявления нарушений ритма и проводимости на каждом этапе хирургической коррекции всем детям проводилось обследование, включавшее: ультразвуковое исследование сердца, пульсоксиметрию, зондирование полостей сердца и ангиопульмонографию, электрокардиографию (ЭКГ), суточное мониторирование ЭКГ до и после оперативного вмешательства.

Результаты. Свобода от нарушений ритма гемодинамической коррекции составила 98,1% (I этап), 80% (II этап), 48,6% (III этап), 41,5% (IV этап). Начиная со II этапа гемодинамической коррекции, наиболее частым нарушением ритма сердца (НРС) была дисфункция синусового узла. При проведении статистического анализа показано, что в группе детей с НРС по сравнению с детьми без аритмий отмечалось значимое снижение сатурации кислорода в капиллярной крови ((73 ± 12)% и (81 ± 4)% соответственно, $p < 0,05$) и повышение среднего давления в легочной артерии после проведения операции тотального кавапульмонального соединения (ТКПС) ((9,6 ± 4,2)% и (11,5 ± 5,8)% соответственно, $p < 0,05$). Средний возраст пациентов с аритмиями в группе пациентов после ТКПС и, соответственно после закрытия фенестрации, был статистически значимо выше, чем в группе детей без аритмий ((72 ± 43) мес и (52 ± 27) мес соответственно, $p < 0,05$). Исследование аритмий у пациентов с функционально единственным желудочком сердца требует проспективного наблюдения, поскольку они существенно влияют на прогноз заболевания и качество жизни.

Ключевые слова: послеоперационные аритмии, дети, единственный желудочек сердца, этапы гемодинамической коррекция ВПС.

КОНФЛИКТ ИНТЕРЕСОВ

Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

ИСТОЧНИК ФИНАНСИРОВАНИЯ

Авторы заявляют об отсутствии финансирования при проведении исследования.

СООТВЕТСТВИЕ ПРИНЦИПАМ ЭТИКИ

Исследование одобрено локальным комитетом по этике НИИ кардиологии ТНИМЦ РАН (г. Томск).

Поступила в редакцию 22.12.2017
Подписана в печать 24.04.2018

Плотникова Ирина Владимировна, д-р мед. наук, руководитель отделения детской кардиологии, НИИ кардиологии, ТНИМЦ РАН, г. Томск.

Джаффарова Ольга Юрьевна, канд. мед. наук, ст. науч. сотрудник, отделение детской кардиологии, НИИ кардиологии, ТНИМЦ РАН, г. Томск.

Свинцова Лилия Ивановна, канд. мед. наук, вед. науч. сотрудник, отделение детской кардиологии, НИИ кардиологии, ТНИМЦ РАН, г. Томск.

Саушкин Виктор Вячеславович, канд. мед. наук, ст. науч. сотрудник, лаборатория радионуклидных методов исследования, НИИ кардиологии, ТНИМЦ РАН, г. Томск.

Ковалев Игорь Александрович, д-р мед. наук, профессор, зам. главного врача, НИКИ педиатрии им. академика Е.Ю. Вельтищева, РНИМУ им. Н.И. Пирогова, г. Москва.

(✉) **Джаффарова Ольга Юрьевна**, e-mail: oyd@cardio-tomsk.ru.