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ANALYSIS OF THE IMMEDIATE RESULTS HEMODYNAMIC CORRECTION ON PATIENTS WITH THE PHYSIOLOGY OF SINGLE VENTRICLE

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Introduction. The single ventricle (SV) is a congenital heart disease characterized by the absence of a septum between the ventricles of the heart. Mortality in the first year of life from reaches more than 35% in the early neonatal period, 65% of children die during the first 6 months of life. In this connection, the issue of studying mortality and ways to reduce it in this defect is very relevant.

Material and methods. In RSSPMS for surgery named. acad. V.Vakhidov for the period from January 2004 to December 2021 diagnostic interventions were performed in 81 patients. Males were 43 (54.4%) patients, female 36 (45.6%). In our series of 81 observations, all patients underwent standard diagnostic methods, including echocardiography, angiocardiography, and multislice computed tomography. Of 81 patients, 60 (74.1%) patients underwent various staged hemodynamic interventions: 7 (8.6%) patients underwent modified BT shunt; 51 (62.9%) patients underwent the bidirectional cavopulmonary anastomosis; 2 (2.4%) patients underwent the Fontan operation.

Results. All the results obtained during the operations were divided into good, satisfactory and unsatisfactory, as well as fatal outcomes. Good results were noted in 20 (33.3%) cases, satisfactory in 24 (40.0%) cases, unsatisfactory results were noted in 3 (5.0%) cases out of 60 operated patients. The cumulative mortality in the study series of patients for the entire period of the retrospective study was 21.6% (13 people).

Conclusion. We noted that for the period 2005-2012, mortality in relation to the number of observations for that period was high, which, in our opinion, is associated with the period of development of methodological approaches in the diagnosis and surgical treatment of this heart disease, however over the period from 2013 to 2021, due to the revision of tactical aspects and the improvement of surgical techniques, mortality decreased by 4.5 times compared to the previous period.

Keywords: physiology of the single ventricle of the heart, hemodynamic correction, surgical treatment, mortality analysis.

Introduction.

The single ventricle (SV) - is a congenital heart disease characterized by the absence of a septum between the ventricles of the heart. The main sign is the

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communication of both atria through the mitral and tricuspid valves with a common ventricle.

Congenital malformations rank third in the structure of infant mortality, half of the deaths of which are congenital heart disease (CHD). The frequency of occurrence of a functionally single ventricle among all congenital heart diseases is 2.5%, among critical ones it is 5.5% per 1000 newborns [13]. Mortality in the first year of life from CHD reaches more than 35% in the early neonatal period, 65% of children die during the first 6 months of life [3]. The natural course of CHD with univentricular hemodynamics is characterized by 70% mortality within 16 years with a well-formed left ventricle, 50% mortality within 4 years with a single right ventricle of the heart [10, 12].

The natural course of SV without surgical correction depends on the anatomical components of this heart disease and its inherent complications: pulmonary hypertension, AV valve insufficiency, hypoxemia, heart failure [8]. The results of our series of observations show that the long course of the defect leads to progressive hypoxemia and a decrease in ejection fraction with the development of congestion in the pulmonary and systemic circulation. The cornerstone in relation to the tactics of treating patients with SV is the intact functional state of the pulmonary circulation, i.e. the absence of severe pulmonary hypertension, which requires targeted clinical research in each specific case. Due to the so-called. "mixing of arterial and venous blood in the common chamber" characteristic of all types of SV [4], the key factor in choosing treatment tactics is the presence of pulmonary artery stenosis and its magnitude, as factors determining the preservation of the functional state of the small circle, since, unlike conventional septal defects, saturation indicators blood do not allow to unambiguously judge the state of the small circle and, as a result, in clinical practice I can be characterized by a paradox [6] In particular, in patients without pulmonary stenosis or with unexpressed stenosis, saturation may remain low, which may be due to both a massive volume of blood mixing at the level of the atria and ventricles of the heart, and a reduced capacity of the pulmonary circulation in an inoperable group of patients due to high pulmonary hypertension.

If with regard to anatomical correction, the criteria for selecting candidates for anatomical correction of the defect are extremely clear, then with regard to hemodynamic correction in SV, everything is not so clear [5].

In this regard, in patients with SV according to the European guidelines and other recommendations in patients with a saturation of 70% and below [1,2], it is an indication for a staged surgical hemodynamic correction of the defect, provided that a functionally acceptable state of the pulmonary circulation is maintained.

With regard to the degree of preservation of the functional state of the small circle during hemodynamic correction of the SV, there are certain limitations in the choice of the stage of surgical correction compared to patients with septal heart defects, which are

regulated individually for the SV in relation to the choice of each individual stage of hemodynamic correction [7].

Among the absolute contraindications to surgical correction of SV are irreversible pulmonary hypertension stage III-IV, as a rule, these are patients older than 3 years, as well as sub- and decompensated heart failure.

Material and methods.

In RSSPMS for surgery named. acad. V.Vakhidov for the period from January 2004 to December 2021 diagnostic interventions were performed in 81 patients, 60 (74.1%) patients underwent various staged hemodynamic interventions: 7 (8.6%) patients underwent modified BT shunt; 51 (62.9%) patients underwent the bidirectional cavopulmonary anastomosis; 2 (2.4%) patients underwent the Fontan operation.

The study of patients with functionally single ventricle with the aim of establishing a diagnosis and identifying concomitant anomalies in the development of the cardiovascular system included general clinical and instrumental research methods: history taking, examination, anthropometry, auscultation, thoracic X-ray, electrocardiography, transthoracic echocardiography. In complex anatomical variants of functionally single ventricle with concomitant CHD, angiocardiography with catheterization of the heart cavities, as well as multislice spiral computed tomography, were performed to assess the severity of intracardiac hemodynamic disorders.

The age of patients at the time of inclusion in the study ranged from 5 months. up to 25 years (average 8.2 ± 6.7 years). The weight of the patients varied from 6 kg to 99 kg (average 23.6±16.9 kg). The body surface area (BSA) ranged from 0.34 m2 to 2.94 m2 (0.85 ± 0.43 m2 on average). Males were 43 (54.4%) patients, female 36 (45.6%) with a sex ratio (1.19:1) presented in Table 1.

Table 1. Distribution of patients by age and sex.

Sex	**Age (years)**			**Total n (%)**
	0-4	**4-12**	**12-30**	
Males	16	18	11	**43 (54,4)**
Females	15	12	9	**36 (45,6)**
Total n (%)	**31 (38,0)**	**30 (36,7)**	**20 (25,3)**	**81 (100)**

The totality of concomitant cardiac malformations with anatomical variants of physiology SV in patients was detected by radiation methods of research with the final intraoperative verification of the results of which are shown in Table 2.

Table 2. Spectrum of concomitant anomalies in SV.

Variants of physiology SV	**Associated malformations**	**n**	**%**
Tricuspid valve atresia	Malposition of great arterias + Pulmonary artery stenosis (PS)	12	15,1
Double inlet SV	TGA + PS	17	21,5

Double inlet SV	PS + anomalous confluence SVC or IVC	4	5,0
Tricuspid valve atresia	Completed AVC + Severe pulmonary hypertension (PH)	2	2,5
Tricuspid valve atresia	ASD + VSD + PS	16	20,2
DORV	TGA + PS	3	3,8
Double inlet SV	Malposition of great arterias + Severe PH	5	6,3
Mitral valve atresia	Malposition of great arterias + PS	3	3,8
Tricuspid valve atresia	TGA +CoA+PDA+ Severe PH	1	1,2
DORV	PS	3	3,8
Ebstein's anomaly: type B / type C by A.Carpentier	No associated anomalies	6	7,6
Tetralogy of Fallot. Hypoplasia of TV and RV.	No associated anomalies	2	2,5
ПФ АВК	Pulmonary artery atresia I type by J.Somerville + PDA + Additional SVC	2	2,5
Corrected TGA. Severe PH	No associated anomalies	1	1,2
Corrected TGA	PS	2	2,5
Total:		**81**	**100**

The analysis of the immediate results of surgical treatment was carried out on the basis of a comparison of subjective and objective indicators of the general condition of patients immediately after surgery and at discharge from the clinic. The study of these indicators was carried out on the basis of inpatient examination of patients in the postoperative period in order to objectively assess their current somatic condition.
For an objective assessment of the general condition of patients, they were assigned a functional class according to the NYHA classification.

The condition of the operated patients was assessed by standard division of the result into good, satisfactory and unsatisfactory, and lethality rates were analyzed separately.

As a rule, today the full cycle of surgical treatment of SV consists of three stages: 1) Modified Blalock -Taussig shunt (MBTS) in order to prepare the capacity of the pulmonary-arterial bed for the upcoming bidirectional cavopulmonary anastomosis; 2) Bidirectional cavopulmonary shunts (BCPS) Glenn operation in order to level the effects of hemodynamic transfer from active (pulsating) to passive (laminar) blood flow in the pulmonary circulation; 3) Complete cavopulmonary anastomosis (Fontaine operation) in order to completely exclude tissue hypoxia during the surgical treatment of patients with SV.

In cases of non-compliance of patients with the criteria for hemodynamic correction and in the presence of severe pulmonary hypertension, patients were discharged without surgery with medication prescriptions for further observation.

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All surgeries for applying MBTS were performed from the thoracotomy approach, and the surgeries for bidirectional cavopulmonary anastomosis of the Fontan operation were performed under conditions of hypothermic cardiopulmonary bypass with pharmaco-cold cardioplegia (CP). General indicators of bypass, CP and temperature regime during BCPS and Fontan operations in patients with SV are presented in Table 3.

Table 3
General indicators of bypass, CP and temperature conditions in operations of BCPS and Fontan in patients with SV

Cardiopulmonary bypass indicators	**(n-60)**
Total cardiopulmonary bypass time (min)	76,7 ± 56,9
Total cardioplegia time (min)	24,9 ± 25,2
Average perfusion temperature (C°)	34,4 ± 2,3

In our study, MBTS was performed in 7 (8.8%) patients. The reason for the imposition of the MBTS was the low saturation, the index Nakata and the index McGoon. Bidirectional cavopulmonary anastomosis was performed in 51 (62.9%) patients. In view of the lack of experience in performing such operations until 2012, BCPS was applied to patients without clear indications and contraindications, this period was characterized by a small number of such interventions and amounted to less than 14% of all cases of BCPS performed in subsequent years, which will be covered in more detail in the section of this chapter devoted to analysis of hospital mortality.

However, after 2012, in the overwhelming majority of cases, when setting indications for surgical treatment according to the Glenn method, we were guided by the criterion of A. Choussat [9].

A complete cavopulmonary anastomosis was performed in 2 (2.4%) patients. All operations of this type were carried out by us in 2012, in this regard, due to the lack of tactical and technical experience in carrying out this type of operation, they were performed without clear indications and contraindications, as a result of which this period was characterized by a small number of such interventions and total mortality.

Results and discussion

In the short term after the operation, the functional state was studied in all 60 (74.1%) operated patients out of 81 examined. The results were evaluated according to the division into the following groups: good, satisfactory and unsatisfactory.

Patients with good results showed a significant improvement in subjective state, positive dynamics of echocardiography data: 1) absence of visual stenosis at the level of interarterial or cavopulmonary shunt; 2) laminar flow on cavopulmonary shunt or pressure gradient (PG) up to 0.8 mm Hg; 3) volemic unloading of the SV in bidirectional cavopulmonary shunts determined by the degree of reduction of the IVC from 40 to 50% in relation to its diameter during the cardiac and respiratory cycle; 4) an increase in saturation by an average of 15 points compared with the initial value, both with BCPS

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and with MBTS; 5) improvement in NYHA functional class by 2 orders of magnitude or more at the time of hospital discharge [11].

The group with satisfactory results included patients according to the following criteria: 1) absence of visual stenosis at the level of interarterial or cavopulmonary shunt; 2) laminar flow on cavopulmonary shunt or PG up to 0.8- 1.5 mm Hg; 3) volemic unloading of the SV in bidirectional cavopulmonary shunts determined by the degree of reduction of the IVC from 30 to 40% in relation to its diameter during the cardiac and respiratory cycle; 4) an increase in saturation by an average of 10 points compared with the initial value, both with BCPS and with MBTS; 5) improvement in NYHA functional class by 1 orders of magnitude or more at the time of hospital discharge.

The group with unsatisfactory results included patients according to the following criteria: 1) the presence of visual stenosis at the level of an interarterial or cavopulmonary shunt; 2) flow on BCPS with a permissible PG from 1.5 mm Hg. and higher; 3) volemic unloading of the SV in BCPS determined by the degree of IVC contraction of less than 30% in relation to its diameter during the cardiac and respiratory cycle; 4) no changes or a decrease in saturation up to 5 points compared with the initial value, both with BCPS and with MBTS; 5) No change or decrease in NYHA functional class at the time of hospital discharge.

Cases of lethal outcomes were included in a separate group and analyzed separately.

Thus, good results were noted in 20 (33.3%) patients out of 60 operated, satisfactory - in 24 (40.0%) and unsatisfactory - in 3 (5.0%) patients. Group data are presented in Table 4.

Table 4
Immediate results of surgical treatment (n=47)

Indicator	**Good n-20**	**Satisfactory n-24**	**Unsatisfactory n-3**	**P ***
Average age (years)	5,5±5,7	9,9±7,4	4,5±0,7	>0,05
Saturation difference (%)	14,6±9,0	12,9±7,5	-7,5±2,1	>0,05
IVC contractility (%)	50,0±0,5	43,3±5,8	-	<0,05
Artificial lung ventilation duration (hours)	8,3±3,4	11,0±6,2	6,3±0,4	<0,05
Carditonics (hours)	29,3±22,4	41,4±32,1	-	<0,05
Stay in the ICU (days.)	1,6±0,9	2,3±1,7	1,0±0,1	<0,05
Post operating period (days.)	11,4±5,9	14,0±8,0	8,0±0,2	>0,05
*- Comparison of subgroups of patients with good and satisfactory results				

Good results were observed in 20 out of 60 patients operated on for SV, which accounted for 33.3% of those operated. The average age of patients in this group was (5.5±5.7 years). By the time of surgery: 6 (10.0%) patients belonged to functional class II; 13 (21.6%) patients to functional class III; and 1 (1.7%) patients to functional class

IV according to NYHA. By the time of discharge, the patients showed a significant improvement in their general condition, the absence of shortness of breath, palpitations. In this group, in 18 (30.0%) cases, BCPS was applied, and in one patient, the operation was performed on an emergency basis for health reasons, in 2 (3.3%) cases, MBTS was applied.

All patients with BCPS at the time of discharge, according to the control postoperative echocardiography, had a laminar flow on the anastomosis, and patients with MBTS had good anastomotic function. The duration of artificial lung ventilation in the postoperative period in this group averaged 8.3±3.4 hours. Cardiotonic support was 29.3±22.4 hours.

All patients in this group showed a significant increase in saturation from 5 to 37% compared with baseline values.

Despite the earlier extubation in this group of patients compared to other patients, the average stay was 1.6 ± 0.9 days, which was due to ongoing cardiotonic support, including in the ward after transfer from intensive care.

With the normalization of intracardiac hemodynamics and general condition, patients of this group after surgery in a satisfactory condition were discharged from the hospital on an average of 11.4±5.9 days for outpatient treatment. Such a wide range of values is due to a longer selection of anticoagulant therapy in some patients, with the aim of achieving indicators of 45-60%, which corresponds to 2.0-2.5 INR.

Satisfactory results were noted in 24 cases out of 60 operated patients for SV, which accounted for 40.0% of operated patients. The average age of patients in this group was (9.9±7.4 years). By the time of surgery: 7 (29.2%) patients belonged to functional class II; 17 (70.8%) patients functional class III by NYHA. By the time of discharge, the patients showed a significant improvement in their general condition, the absence of shortness of breath, palpitations. In this group, BCPS was applied in 22 (91.6%) cases, MBTS was applied in 2 (8.3%) cases.

In all patients with BCPS at the time of discharge, according to the control postoperative echocardiography, a laminar flow or an acceptable pressure gradient in the BCPS area from 0.8 to 1.5 mm Hg was noted, and in patients with MBTS, the anastomosis function was assessed as satisfactory. The duration of artificial lung ventilation in the postoperative period in this group averaged 11.0±6.2 hours. It should be noted that patients in this subgroup required significantly longer cardiotonic support and amounted to 41.4±32.1 hours.

All patients in this group also showed an increase in saturation from 3 to 33% compared with baseline values, and the average increase in saturation was 12.9±7.5%.

Despite the desire for earlier extubation in this group of patients, in order to ensure earlier suction action of the chest during the act of breathing, nevertheless, the average residence time was 2.3 ± 1.7 days, which in turn was due to a longer course of heart failure, which required longer cardiotonic support in intensive care.

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Unsatisfactory results were noted in 3 cases out of 60 operated patients for SV, which accounted for 40.0% of operated patients. The average age of patients in this group was (4.5±0.7 years). By the time of surgery, all patients were in functional class III by NYHA. By the time of discharge, the patients showed a significant improvement in their general condition, the absence of shortness of breath, palpitations. In this group, BCPS was applied in 1 (91.6%) case, MBTS was applied in 2 (8.3%) cases.

Despite the somatically stable course of patients in this group and the rapid transfer from the intensive care unit to the department, the criterion for including patients in this subgroup was a decrease in saturation compared to the initial value at the time of discharge from 5 to 10% in patients after MBTS, as well as the presence of BCPS of significant regurgitation on AV valves up to 2-3 degree. The duration of artificial lung ventilation in the postoperative period in this group averaged 6.3±0.4 hours, and cardiotonic support was required only in one case for a short period of up to 60 minutes.

All patients in this group also showed an increase in saturation from 3 to 33% compared with baseline values, and the average increase in saturation was 12.9±7.5%.

Postoperative complications are presented in table 5 and were noted in patients in the group with satisfactory, as well as in the group with a fatal outcome.

Table 5. Complications of the postoperative period

The nature of postoperative complications	**n**	**%**
Satisfactory group results	**7**	**11,6%**
Delayed healing of p / o wounds	5	8,3%
Pulmonary complications (pneumonia)	2	3,3%
The lethal group	**13**	**21,6%**
Progressive acute heart failure	13	21,6%
Acute respiratory failure	4	6,6%
Acute trombosis of BCPS	4	6,6%
Gangrene of the upper limb	1	1,6%
Pulmonary hypertension crisis	1	1,6%
Syndrome of disseminated intravascular coagulation	1	1,6%
Multiple organ failure	1	1,6%
TOTAL:	**20**	**33,3%**

In two cases, patients required repeated surgical interventions: in one case due to superior vena cava syndrome, and in another case due to intrathoracic bleeding after MBTS.

Our series of observations dates back to 2004 to the present, therefore, in the process of analyzing hospital mortality, our attention was drawn to the fact that a significant increase in the number of both diagnostic and surgical interventions has been observed from 2013 to the present. While from 2004 to 2012 inclusive the number of diagnostic and treatment interventions is significantly lower compared to the same period from 2013 to 2021, at the same time, despite a considerable number of

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observations during this period, the number of refusals from operations is relatively high, and the surgical activity in the treatment of this nosology is low, which is due both to the late diagnosis of the disease and the lack of comprehensive knowledge and surgical skills in the treatment of SV. Statistical data on diagnostic and treatment interventions and mortality for the entire observation period is presented in Table 6.

The years of observation in 2012 and 2020 deserve special attention, as they are characterized by the complete absence of SV cases both in diagnostic and surgical terms. The lack of observations for these periods is due to the reorganizational aspects of our institution. In 2020, the number of SV observations is not available due to the global Covid 19 pandemic and the introduction of restrictive quarantine measures throughout the country.

At the same time, from 2013 to the present, there has been an inverse trend, both the overall detection rate of the defect and surgical activity have increased, while there is a steady trend in a multiple decrease in mortality in relation to the total number of operated patients over the specified observation period.

Table 6. Chronological structure of surgical activity and mortality

Years	**Non operated patients**		**Operated patients**		**Dead patients**		**TOTAL**	
	n	**%**	**n**	**%**	**n**	**%**	**n**	**%**
2004	-	-	1	1,2%	-	-	1	1,2%
2005	5	6,2%	3	3,7%	1	1,2%	8	9,9%
2006	2	2,5%	2	2,5%	2	2,5%	4	4,9%
2007	5	6,2%	-	-	-		5	6,2%
2008	-	-	1	1,2%	1	1,2%	1	1,2%
2009	1	1,2%	2	2,5%	1	1,2%	3	3,7%
2010	3	3,7%	1	1,2%	1	1,2%	4	4,9%
2011	-	-	2	2,5%	1	1,2%	2	2,5%
2012*	-	-	-	-	-	-	-	-
2013	-	-	4	4,9%	-	-	4	4,9%
2014	-	-	4	4,9%	-	-	4	4,9%
2015	-	-	1	1,2%	-	-	1	1,2%
2016	-	-	5	6,2%	1	1,2%	5	6,2%
2017	3	3,7%	14	17,3%	3	3,7%	17	21,0%
2018	2	2,5%	12	14,8%	2	2,5%	14	17,3%
2019	-	-	7	8,6%	-	-	7	8,6%
2020**	-	-	-	-	-		-	-
2021	-	-	1	1,2%	-	-	1	1,2%
TOTAL:	**21**	**25,9%**	**60**	**74,1%**	**13**	**16%**	**81**	**100%**

***** - the absence of patients during this period is due to the launch of the cardiocenter.
****** - the absence of patients during this period is due to the global coronavirus pandemic.

Thus, in the process of comparative analysis, we conditionally divided our series of observations into two periods: the period from 2005 to 2012. and the period from 2013 to the present presented in table 7.

Table 7 Chronological structure of postoperative mortality

Time periods	**Observed patients for the period**		**Dead patients during the period**		
	n *	%*	n *	%*	%**
2005-2012гг. *	12	20,0	7	58,3	11,6
2013-2021гг.*	48	80,0	6	12,5	10,0
TOTAL: n (%)**	**60**	**100,0**	**13**		**21,6**
* - the percentage of patients in relation to the number of observations for the specified period. **- percentage of patients in relation to the total number of observations for both periods					

The cumulative mortality in the study series of patients over the entire period of the retrospective study was 21.6% (13 people) of all operated patients, however, when analyzing each period separately, we noted that for the period 2005-2012, despite the small number of observations, mortality in relation to the number of observations for that period was high, which, in our opinion, is associated with the period of development of methodological approaches in the diagnosis and surgical treatment of this heart disease.

This period is represented by a full range of operations performed in patients with SV from the imposition of MBTS to the Fontan operation, however, due to the lack of comprehensive knowledge and methodological approaches to assessing patients and setting correct indications for surgical treatment, this period is characterized by an extremely high mortality rate of up to 58.3%.

In relation to the next period of 2013-2021, there is an increase in the total number of observations, which indicates an improvement in the detection of the defect, which, in our opinion, is due to the rapid development of diagnostic systems of that period, in particular, with the advent of new generation echocardiographs, as well as the introduction of diagnostic MSCT.

Thus, with the development of diagnostic systems, the tactical and technical aspects of surgical treatment and the improvement of the material and technical base as a whole, there was an increase in both the detection rates of the defect and a multiple decrease in mortality compared to the previous period by 4.5 times.

When analyzing the causes of mortality in most cases, among the main factors were acute cardiovascular failure in 13 (21.6%) cases and acute respiratory failure with acute thrombosis of BCPS in 4 (6.6%) cases, respectively.

All of the deaths described above occurred between 2005 and 2012. when there were no clear criteria for selecting candidates for this type of surgical intervention in the clinical work.

Lethal cases that occurred in the period from 2013 to 2021, in our opinion, are mainly associated not so much with the tactical as with the technical aspects of surgical treatment, so many diagnostic and tactical aspects have been significantly improved on the basis of our department.

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When analyzing mortality rates for the presence of factors affecting the outcome, we established the following relationships:

With regard to the factors affecting the postoperative outcome, we noted a clear relationship between the time of extubation and mortality. Paradoxically from the point of view of resuscitation, the fact that the longer the patient is on prolonged mechanical ventilation, the higher the likelihood of death. In the group with good results, the average time spent on mechanical ventilation was 8.3 hours, while the lengthening of this indicator by more than 7 times was noted in the group with a fatal outcome with $p<0.05$.

Among the diagnostic methods, we have established the relationship between the size of the cardiothoracic index index and the lethal outcome in patients with cardiothoracic index more than 60% $p<0.05$. It is noteworthy that we did not find such a statistical relationship with respect to the end diastolic volume of SV, which often determines the size of the heart shadow.

It was noted that the combination of SV with an open ductus arteriosus in terms of prognosis may be unfavorable for the reason that a similar variant was noted in our observations in two cases. Despite the fact that in both cases, in the process of applying BCPS, in both cases, the PDA was ligated, nevertheless, in both patients, a lethal outcome was noted. However, this judgment is of a hypothetical nature, since, due to the small number of both patients with SV and its combination with PDA in our series of observations, it did not allow us to conduct a correlation analysis for this symptom.

We also noted the prevalence among patients with a fatal outcome of 4 cases of patients with Ebstein's anomaly, which, in our opinion, is due to the need for intraoperative correction of regurgitation of tricuspid valve. This thesis is indirectly confirmed by the Choussat's criteria, however, on the scale of the current study, due to the small number of observations on this trait, it is not possible to identify statistically significant relationships.

Conclusion

The analyzing the data for the presence of predictive indicators affecting the outcome of surgical treatment, we established the following relationships: 1) there is a clear relationship between extubation time and mortality with a significance of $p<0.05$. which boils down to the following regularity, the longer the patient with SV after BCPS stays on mechanical ventilation, the higher the mortality. Such a paradox is due to the absence of the "thoracic pump" function, which facilitates the work of BCPS due to negative pressure in the chest during spontaneous breathing; 2) There is a statistically significant relationship between cardiothoracic index size and mortality with a borderline value of 60% with statistical significance $p<0.05$. Among patients with cardiothoracic index more than 60% mortality is higher than in patients with a value below this value. At the same time, it is noteworthy that in relation to the end diastolic volume of SV, which often determines the size of the heart shadow, we did not find such a statistical relationship; 3) there are prerequisites to assert that SV in combination with

open ductus arteriosus can be unfavorable in terms of prognosis, however, due to the small number of both patients with SV and its combination with PDA in our series of observations, it does not allow us to state this statistically reliably, but this thesis also confirmed by European guidelines and other clinical guidelines; 4) With regard to marginal forms of Ebstein's anomaly, there is a high probability of a relationship with a fatal outcome, however, due to the small number of observations on this trait, it is difficult to prove with a statistically significant degree of certainty. However, this thesis is indirectly confirmed by Choussat's criteria mentioned above in this paper.

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