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RESULTS OF CORRECTION OF PARTIAL ANOMALOUS PULMONARY VEIN DRAINAGE

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Abstract:

The paper presents the materials of patients with partial anomalous pulmonary drainage operated on from 2001 to 2021 in the Department of Congenital Heart Disease Surgery of V.Vakhidov RSNPMCh, the age of patients was 1,4-45 years. The diagnosis of the defect was based on the analysis of the combined data of clinical and special methods of investigation. Surgical correction of malformation was performed by median sternotomy and right lateral thoracotomy, with artificial circulation. Peculiarities of the course of the postoperative period depending on the type of malformation were performed. Changes in cardiac parameters were analyzed using electrocardiography, echocardiography and roentgenography.

Keywords: partial anomal perfusion of pulmonary veins, surgical treatment, pulmonary hypertension, evaluation of results

RELEVANCE. The problem of congenital heart disease (CHD) is one of the most significant for children's health care due to the high early mortality rate, propensity to form severe, chronic complications and the development of irreversible changes in the body. On average, CHD accounts for about 35% of all congenital malformations. At the current stage of development of the national healthcare system, many measures are taken to improve the results of treatment of patients with CHD by introducing modern principles of prevention, conservative and surgical treatment. Abnormal pulmonary vein drainage (APLV) is a rare and complicated congenital heart defect characterized by the insertion of one or more pulmonary veins into the right atrium or large circulatory system [1,4,14,16]. ADLV is associated with atrial septal defect (ASD) in 90% of cases. Depending on the number of pulmonary veins inflow, there are hourly ADLV (CHADLV), when one or several, but not all pulmonary veins inflow to the right side, and total ADLV, when all pulmonary veins inflow to the right side of the heart. There are several anatomic variants of the defect. The most accepted classification is that of R. Darling et al. [11], who consider the forms of malformation depending on the level of connection of pulmonary veins with the system of the great circle of circulation. A distinction is made between: 1) supracardiac form (type I), in which all pulmonary veins are drained by a common collector into the superior or supplementary left superior vena cava; 2) cardiac form (type II), in which the mouths of pulmonary veins open into the right atrium or coronary sinus; 3) infradiaphragmatic form (type III) with pulmonary veins draining into portal or inferior vena cava system; 4) mixed form (type IV) - pulmonary veins flow separately into one of the above formations. According to different authors, ADLV occurs from 0.7 to 9% of all congenital heart defects [3,4,6,10,13,15], and almost 80% of these patients have the partial form [3,7,14]. Single works [5,8,9,12,15] analyze the results of ADLV surgical treatment, the issues of rehabilitation of operated patients and their rational labor management, and the evaluation of distant correction outcomes often uses methods that do not allow performing an objective quantitative analysis of the occurring changes in hemodynamics, their regression [2,17]. In conclusion, it should be noted that the literature analysis indicates that the issues of studying the results of malformation correction are topical. Therefore, the development of this problem is of great practical and scientific importance.

OBJECTIVE: to study the peculiarities of postoperative period and analyze the results of surgical treatment of ChADLV depending on the type of malformation.

Material and methods: 149 patients were operated for partial anomalous pulmonary vein drainage from 2001 to 2021 in V.V. Vakhidov RSNPMC.

149 patients with different anatomical variants were operated on. Their age ranged from 1.4 to 45 years, averaging 16.2±0.1 years. There were 62 (41.6%) male and 87 (58.4%) female patients (Table 1).

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Table 1.
Distribution of patients with CHADLV according to sex and age

sex	Age, years						total
	≤ 3	4-7	8-15	16-20	21-30	>30	
man	6	15	19	14	5	3	62
woman	6	12	25	11	16	17	87
total	12	27	44	25	21	20	149

All patients with CHADLV were divided into the following main types (Table 2).

Table 2.
Characteristics of patients with PDLV depending on the type of malformation

Indicator	CHADLV type				Average
	I	II	III	IV	
Age, years	15,9	16,9	18,9	13,4	16,2
Body weight, kg	40,7	39,3	46	31,2	40,4
Height, cm	144,4	142,1	136,2	139,3	143,5
PPT m2	1,51	1,38	1,72	1,91	1,25
Erythrocytes, x10*13/l	4,5	4,7	5,1	5,2	4,8
Hemoglobin, g/l	120	118	122	128	122
Capillary RO2, mm Hg	97	100	102	98	99,2
HBO2, %	90	92	94	89	91,2

All patients with CHADLV underwent a complex examination, which included: anamnestic data collection; general clinical methods, including laboratory methods (general and biochemical blood tests, hemostasis system tests, general urine tests, blood gas composition); electrocardiography in standard and chest leads, chest X-rays; complex echocardiographic examination. Clinical manifestations of volumetric overload of the right ventricle consisted of the symptoms of heart failure and arterial hypoxemia. Surgical tactics and the volume of surgical interventions performed in CHADLV depended on the form of malformation (Table 3). The largest contingent - 83 (55.7%) of patients with CHADLV were of supracardiac type. The nature and level of LV drainage, as well as the presence of concomitant defect were of great importance in choosing one or another method of malformation correction and its provision. In all cases, the correction included LV orifice dislocation into the left atrium using ASD plasty as well as elimination of concomitant heart defects. All types of malformation correction were performed with artificial circulation. As a rule (86.5%), "open" heart operations were performed via median sternotomy, and in 20 (13.4%) cases right lateral thoracotomy was used. Aortic and vena cava cannulation was carried out according to the standard protocol: in 83 cases with I-type ChADLV in VPV the latter was cannulated with the "L"-shaped cannula above the site of pulmonary veins inflow. In 4 (2.6%) cases, LV dislocation was performed by simple suturing of the free lateral edge of the ASD to the PP wall to the right and above the mouth of abnormally flowing LVs; ASF plasty with auto- or xenocardial patch with simultaneous dislocation of anomalous accessory LV mouths into LV through ASF was performed in 54 (36,2%) cases; in 83 (55,7%) patients with supracardiac and in 6 (4,02%) patients with mixed type of ChADLV we performed surgery according to J. Kirklin;

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two patients underwent anastomosis between the common manifold of all LVs and the LV auricle.

Table 3.
Distribution of patients with ChADLV according to the type of surgical intervention

Type of surgical intervention interventions	Number of patients	CHADLV type			
		IType	II Type	IIIType	IYType
	4	-	4	-	-
Septoplasty	54	-	54	-	-
ASD surgery with simultaneous dislocation of LV ostium into LV cavity	75	68	-	1	7
LV	2	2	-	-	-
Surgery according to J.Kirklin	7	7	-	-	-
Anastomosis between the LV collector	6	6	-	-	-
and LV auricle	149	83	58	1	7

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The open heart in 54(36.2%) cases was provided with normothermic (46.5±3.4 min) and in 95(63.7%) cases - with hypothermic artificial circulation (68.5±5.6 min) with pharmaco-cold cardioplegia (46.6±3.2 min) (Table 4). Out of 149 patients with CHADLV, 145 patients had an abnormal drainage combined with ASD; in 4 cases there was an open oval window, and in 17 cases the interatrial septum was dissected up or down due to the small size of the defect.

Table 4.
Duration of artificial circulation and cardioplegia

IR with normothermia	IR with hypothermia	Cardioplegia
46.5 + 3.4 min.	68.5 + 5.6 min	46.6 + 3.2 min
n = 54(36.2%)	n=95(63.7%)	n= 128(85,9%)

RESULTS AND DISCUSSION: Analyzing our own material, we tried to assess the adequacy of the operation on the basis of ECG (Table 5). For this purpose, ECG picture was considered before and in different terms after the operation. Thus, indirect signs of right heart hypertrophy after correction of malformation remained in almost all cases, especially with LH. In some cases the above-mentioned was joined by the phenomena of left-sided overload, as a rule, in patients with degree III pulmonary hypertension. Besides, there were cardiac rhythm and conduction disturbances in the postoperative period related to surgical aggression and manipulations near the conduction pathways or the vessels feeding them. Electrocardiography and phonocardiography in the immediate postoperative period showed positive dynamics consisting in decreased degree of hypertrophy and overload of the right heart, disappearance of the characteristic murmur of malformation with decreased accent of II tone over the pulmonary artery.

Table 5.
Comparative quantitative characteristic of ECG findings of patients in pre- and early postoperative periods.

ECG	Before surgery n (%)	After surgery n (%)
SIGNS	137(91,9%)	135(90,6%)
Sinus rhythm	12(8,05%)	14(9,4%)
Various arrhythmias	27(18,1%)	17(11,4%)
Metabolic changes	94(63,08%)	94(63,08%)

Blockade of the right bundle branch (90 incomplete, 4 complete), which was observed in 94 (63,08%) patients before the operation, remained in the nearest postoperative period in all patients. Atrioventricular block of different degree after correction of ADLV occurred in 7 (3,1%) patients,

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though before the operation it wasn't observed. Heart rhythm disturbances (ventricular rhythm, bigemia and rhythm migration), connected with surgical intervention in the nearest period after correction were in 14 (9,4%) patients. After the arterio-venous blood flow is eliminated, the minute volume of the large circulatory system increases and correspondingly, the work of the left heart increases after the operation. Therefore, 36 (16.2%) patients had signs of left heart overload on ECG, which had been absent before the operation.

In the nearest postoperative period according to the clinical data in 183 (82,8%) patients X-ray signs were revealed testifying to a good result of the anomalous pulmonary veins drainage correction. Thus, in this group of patients in the immediate postoperative period there was revealed a significant positive dynamics from the side of the small circle of circulation, i.e. decrease of arterial hypervolemia (Table 6) and roentgenocardiometric parameters (decrease of CPB, heart volume, Moore index).

Table 6.

Quantitative assessment of radiological signs of patients operated on for ADLV.

Degree LH	IAC hypervolemia						Увеличение ПП			
	not		moderate		pronounced		moderate		pronounced	
	A	B	A	B	A	B	A	B	A	B
	1	56	47	12	-	-	33	-	-	-
I	-	-	97	78	-	-	16	66	4	-
II	-	-	4	3	9		4	9	5	-
III	1	56	139	93	9	-	140	75	9	-

Примечание: А - до операции; Б - после операции.

However, the degree of reduction of these cardiometric parameters corresponded to the severity of the defect and the severity of impaired hemodynamics in ICC. Patients with initial LV (Ist) had complete normalization of the mentioned parameters; patients with IIst degree of LV had significant decrease of malformation signs, and patients with IInd degree of pulmonary hypertension had only minor dynamics of cardiometric parameters, despite significant decrease of ICC hypervolemia degree. The increase of the left heart sections was revealed in 80 (53.6%) out of 149 operated patients in the immediate postoperative period that testifies to the elimination of the left-right airflow and the increase of the load on the left heart sections. According to the literature data [13,93,200,228], enough patients operated with ADLV can still have signs of venous stasis in DC due to inadequate size of anastomosis, presence of pulmonary vein stenoses and creation of relatively narrow channel, performing blood outflow from LV to LP. Taking into consideration this fact, we paid special attention to the state of the lung pattern in the nearest postoperative period: only 4 (2.6%) out of 149 operated patients had stasis in the lungs; one of them was operated repeatedly and the other one could be cured by medicamentous treatment. In our opinion, a small number of such complications indicates the adequately performed correction of malformation in patients with ADLV. X-ray examination showed shock lung in one patient after the operation and pneumonia in another patient. Apart from the above mentioned X-ray signs, 14 (9.4%) patients revealed during the first 2-3 days signs of cardiac insufficiency (exudate in pleural cavities, slight congestion in MAC), which were treated with medications. In comparison with preoperative echocardiographic criteria in the immediate postoperative period the following results were obtained. Thus, there was a decrease in linear dimensions of the right heart, disappearance of volume overload; there were signs of left heart enlargement (Fig. 27). Most patients had increased ejection fraction (108% of normal), stroke volume (110% of normal), and correspondingly, cardiac index (3.94 l/min/cm3). Myocardial contractility was within normal range in 73.7% of patients. Doppler study did not reveal any pathological flows; the complete integrity of interatrial septum was established in all patients.

CONCLUSION. Thus, the following immediate results of surgery after ADLV correction were revealed: in 125 (83,8%) - good, in 13 (8,7%) - satisfactory and in 11 (7,3%) - unsatisfactory. There were no in-hospital outcomes. Improvement of anatomical and functional indices was registered in all patients according to ultrasound investigation, ECG and X-ray investigation in the nearest postoperative period. Electrophysiological methods of investigation showed that in the nearest postoperative period arrhythmias occurred in 6.5% of the operated patients. Radiological methods of investigation revealed the disappearance of hypervolemia in the small circle of blood circulation and normalization of the heart size in almost all patients.

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Postoperative echocardiography data prove that in the nearest postoperative period the right heart size decreases and vice versa the left heart size increases; indices of myocardial functional ability increase. We should emphasize that good and satisfactory results were obtained exactly in those patients with preoperative grade I and II pulmonary hypertension and, what is important, the operated were under 18 years old.

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