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# EXPERIENCE OF SURGICAL CORRECTION OF ANOMAL DRAINAGE OF PULMONARY VEINS

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**ABSTRACT**

*Abnormal pulmonary venous drainage (APVD) is a congenital heart disease in which some or all of the pulmonary veins flow into the right atrium, coronary sinus, or vena cava. The incidence of abnormal pulmonary venous drainage ranges from 0.7% to 9% of all CHD according to clinical data [3,8,15].*

**Relevance.** Abnormal pulmonary venous drainage (APVD) is a congenital heart disease in which some or all of the pulmonary veins flow into the right atrium, coronary sinus, or vena cava. The incidence of abnormal pulmonary venous drainage ranges from 0.7% to 9% of all CHD according to clinical data [3,8,15]. If one or more pulmonary veins drain into the right atrium or a large circle, this form of defect is called partial. Partial anomalous pulmonary venous drainage (PAPV) is usually associated with an atrial communication. Less often - the interatrial septum is intact [9,10,12,16] . When the mouths of all pulmonary veins flow into the venous system of the systemic circulation or the right atrium, they speak of complete (total) anomalous pulmonary venous drainage (TADLV). Most often (in 97% of cases), the veins draining from the right lung are abnormally drained [3,4] . With TADLV - all four pulmonary veins from both lungs are connected into one wide collector and this vessel of oxidized arterial blood does not communicate with the left atrium, as it should, but is connected to the venous system of the body, usually through a large vein. Arterial blood, thus, bypassing the heart, enters the large veins and into the right atrium. Only here, having passed through an atrial septal defect (ASD), will it end up where it should be initially - in the left atrium, and then it makes its usual path through the systemic circulation. But children with this defect are born full-term , and the heart copes with such a situation for some time. However , this time can be very short [11,20] . Until now, according to the authors, the surgical correction of ADLV has been basically solved, but, despite such a unity of views on the indications for the correction of such a pathology, many private issues remain the subject of discussion [1,4,6,18]. Basically, they relate to the methods of surgical correction themselves, operative access, the stages of the operation and the conditions for providing the operation. These points, although not of fundamental

importance, but one way or another affect the final result [2,7,9,20]. With this in mind, in this report we present our approach to the surgical treatment of patients with ADLV.

**Purpose of the study:** To evaluate the results of surgical correction of abnormal pulmonary venous drainage.

**Material and methods:** 166 patients aged 3 months and older were operated on at the RSSPTSKh named after academician V.Vakhidov and the cardiosurgical department of the ASMI clinic from 2010 to 2021 . up to 45 years, mean age (11.9±0.7) years, with different anatomical variants of ADLV. The diagnosis of the defect was based on the analysis of the totality of data from clinical and special research methods, including echocardiography, multispiral computed tomography with contrast and catheterization of the heart cavities with angiocardiography . Operations were performed from a median sternotomy, only in 20 (33.2%) cases of operations for this congenital heart disease (CHD) the method of operative access was the right anterolateral thoracotomy, and in 1 (1.6%) case - left-sided thoracotomy. Operations on the "open" heart were performed in 92 patients with normothermic (45.3±2.01) min, and in 74 patients with hypothermic (58.2±1.8) min under cardiopulmonary bypass with pharmaco -cold cardioplegia (Table 1).

Table 1

Duration of cardiopulmonary bypass and cardioplegia

| I K with normothermia | I K with hypothermia | Cardioplegia |
|---|---|---|
| 45.3 ± 2.01 min. | 58.2 ± 1.8 min | 46.6 ± 3.2 min |
| n =92 | n= 74 | n =166 |

**Results of the study:** For the diagnosis of ADLV, radiography was performed, in which one can see the symptom of "Turkish saber, figure eight" [12,17], ECG, ultrasound of the heart, probing of the heart cavities ( angiopulmonography , atrial and ventriculography ), MRI or multislice CT . MRI is recommended when echocardiography is inconclusive [15] . Recently, we often use multislice computed tomography, which can provide information on the topographic anatomy of abnormal pulmonary venous drainage. Patients with unclear RV volume overload were recommended to be hospitalized in specialized institutions in order to conduct further diagnostic studies and identify congenital heart disease.

*The optimal age for performing surgical interventions is 1-2 years of age.* The only method of treatment is surgery, which is carried out under the conditions of IR, hypothermia and pharmacological cold cardioplegia [1,4,6,7,8,17]. There is no other treatment. Method for surgical correction of partial anomalous pulmonary venous drainage is determined by the type of defect: this takes into account the level of drainage, the size and location of the ASD, consists in the formation of a tunnel between the mouths of the right pulmonary veins and the cavity of the left atrium, in order to avoid obstruction of the tunnel (Fig. 1), in some cases, a direct anastomosis of the superior vena cava (SVC) with the right atrial appendage is performed. Depending on the above criteria, numerous options are used for reimplantation of the orifices of the pulmonary veins or their common collectors into the left atrium. To eliminate interatrial communication, suturing or

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plastic ASD is performed.

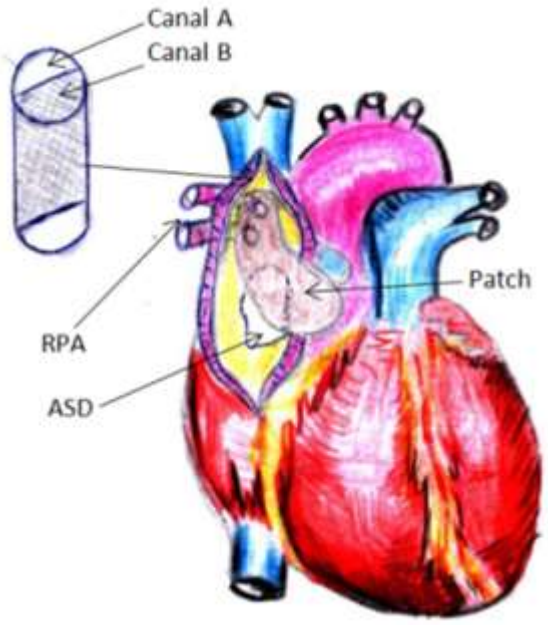


**Fig.1** . Scheme of creating a tunnel in the SVC during correction of the supracardial type of PALV.

Surgical correction of total anomalous drainage of the pulmonary veins is a rather complicated operation, it is performed on the open heart and lies in the fact that it is created anastomosis between the common collector of the pulmonary veins and the left atrium, and the hole in the interatrial septum is closed with a patch, ligation of the pathological communication of the pulmonary veins with venous vessels [3,20] . Thus, after the operation, normal blood circulation is restored in two separated circles . In operated patients, the duration of mechanical ventilation after surgery varied from 1 h 20 min to 18 h 30 min. The time spent in the ICU did not exceed 2 days. Depending on the level of confluence of the pulmonary veins and the location of the ASD, patients were divided into 4 groups: supracardial, cardiac, infracardial and displaced. Of 166 patients, 159 ADLV was combined with an atrial septal defect; 7 patients did not have an ASD and 17 had a small defect, in all these patients the ASD was expanded to the required size so that the blood flowing from the pulmonary veins would not obturate . In 32 patients one pulmonary vein anomalously emptied, in 73 two and in 18 three. Of the 166 patients, 43 had TALV, in 14 cases all pulmonary veins were drained into the superior vena cava, in 27 cases there was a cardiac form and in 2 cases a mixed form of TADLV. In 3 cases, a two-stage (palliative, then radical) TALV correction was performed. Depending on the level of entry of the pulmonary veins and the location of the ASD, the patients were divided into 4 groups: in patients of the first group (29 patients), abnormal pulmonary veins emptied into the SVC; in patients of the second group (114 patients) - into the right atrium; in 5 patients of the third group, the abnormal pulmonary veins flowed into the inferior vena cava (IVC) and in 18 patients the place of confluence was mixed. In 7 (11.6%) operated patients, there was an additional left-sided SVC opening into the coronary sinus. 12 patients underwent Warden operation [21] , including its modification (12 corrections, which accounted for 19.9%). In 87 cases, an atrial septal defect was closed with simultaneous relocation of abnormally draining pulmonary veins, of which, in 11 cases, the technique was supplemented with atriocaval orifice plasty . Postoperative bed-day was 11.4±5.2.

In most cases, the condition of children with complete ADLV turned out to be “critical” from the very beginning of life [3,19,20] . If nothing is done, they will die in a few days or months. An emergency variant of the procedure was used - the expansion of the defect during probing as the first, life-saving stage, which allows you

to somewhat delay the main intervention [19] .

**Discussion:** After surgery, sick sinus syndrome may develop, an increase in pulmonary hypertension associated with inadequate provision of outflow pathways from the pulmonary veins [7,16] . Early postoperative symptoms such as fever, fatigue, vomiting, chest pain or abdominal pain may indicate postcardiotomy syndrome with cardiac tamponade, in which case an immediate echocardiographic study is recommended to rule out this complication [7,13,15] . Lethal outcomes in our group were in 7 patients (11.6%) of all those operated on with ADLV. Endovascular procedures were performed in 4 patients. In three cases, an emergency expansion of the open oval window was performed to save the life of the child. Subsequently, a radical correction of TADLV was carried out. The natural course of total anomalous pulmonary venous drainage is unfavorable: 80% of children die in the first year of life. Patients with partial anomalous pulmonary venous drainage may live to 20 to 30 years of age. The death of patients is associated with severe heart failure or lung infections [15,19] .

**Conclusions. Thus, the results of** surgical correction of ADPV are satisfactory, but intra- and postoperative mortality among newborns remains high. Currently, surgeries for PAPV are performed with good results, characterized by satisfactory outcomes in the immediate and long-term periods after surgery, which was also confirmed by our clinical studies. The choice of surgical correction technique depends directly on the anatomy of the confluence of abnormally draining pulmonary veins.

The use of multislice computed tomography makes it possible to obtain information about the topographic anatomy of abnormal pulmonary venous drainage. Patients with unclear right ventricular volume overload are recommended to be hospitalized in specialized institutions in order to conduct further diagnostic studies and identify congenital heart disease. The diagnosis of ADLV is confirmed by an ACG study with separate injection of a contrast agent into the right and left pulmonary arteries.

The long-term results of the operation are quite good - after all, the main defect has been eliminated. However, children should be under the supervision of cardiologists because complications are possible in the form of rhythm disturbances or narrowing of the pulmonary veins at the suturing sites (this is due to the fact that the heart that has undergone such a major operation continues to grow). And again we want to emphasize: this child is not disabled. He should lead a completely normal life. But we just want to emphasize that children with ADLV need immediate specialized help, which is absolutely real today. It is important to know the basic approaches to the tactics of monitoring patients with ADLV. Before and after surgical treatment, rehabilitation measures should be carried out, taking into account the time elapsed after the intervention and the severity of heart failure. Patients, their parents, and primary care physicians should be instructed to report fever or unusual symptoms (chest or abdominal pain, vomiting, unusual fatigue) in the first weeks after surgery, as these symptoms may represent early signs of cardiac tamponade.

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