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SUCCESSFUL CORRECTION OF PARTIAL ANOMALOUS DRAINAGE OF PULMONARY VEINS INTO THE SUPERIOR VENA CAVA

Mamatov Mukhammadjon Akhmadovich
Butaboev Zhasur Makhmudjonovich.
Department of General Surgery
Andijan State Medical Institute

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Abstract:

This scientific paper describes the authors' experience in the correction of partial anomalous pulmonary vein drainage (PAPVD) in an adult patient. The types of surgery and long-term results in PAPVD in adults are analyzed on the basis of the literature review. The relative rarity of clinical observations of PAPVD in patients over 30 years old is due to the isolated form of malformation without pulmonary vein obstruction and concomitant CHD. The experience with PAPVD correction in adult patients is limited to single surgeries; according to the literature, most adult patients with PAPVD come to the attention of cardiologists and cardiac surgeons after 20 years of age. Therefore, study of clinical course and diagnostic methods of PAPVD in adult patients is relevant to enable timely detection of the defect, rational planning of examination and selection of optimal correction method.

Keywords: congenital heart disease in adults, surgery of partial anomalous pulmonary vein drainage, interatrial septal defect, vena cava, echocardiography, radiography, findings.

INTRODUCTION. Partial abnormal pulmonary vein drainage (PADLV) is a congenital heart defect (CHD) characterized by the insertion of one or more (but not all) pulmonary veins into the right atrium, vena cava or their major tributaries [1,5,7,10,17,]. The finding of this malformation is considered an absolute indication for surgical treatment [9,11,14]. The incidence of PAPVD ranges from 0.9 to 6% of all congenital heart defects [1,3,13,15,16]. Abnormal drainage of the right lung (97.2%) is observed in the majority of cases, and the left lung in the remainder. Literature analysis shows a wide variety of surgical treatments for partial anomalous drainage of the right pulmonary veins into the superior vena cava (AV), which depends on the anatomical components of the malformation, the variety of location of abnormally draining pulmonary veins, and interatrial septal defect (ASD) [2,9,11,13,14]. The average life expectancy of patients with PAPVD does not exceed 37-40 years. At the same time, it is known that the presence of ASD has a significant impact on patients' quality of life [3,5,12]. Given the rarity of PAPVD correction in adults, especially after 30 years of age, we consider it appropriate to present our own clinical observation.

THE AIM IS TO EVALUATE AND ANALYSE he results of correction of partial anomalous pulmonary vein drainage into the superior vena cava using a biplane technique through a systematic review of the literature.

MATERIAL AND METHODS: The most common method of surgical treatment of PAPVD in the VEP is to create a manifold moving abnormally draining pulmonary vein into the left atrium (PL) with subsequent plasty of the anterior wall of the VEP with a patch [10,12,14]. The second method of PAPVD surgical treatment consists in moving abnormally draining pulmonary veins into the LP followed by creation of "end-to-end" anastomosis between the distal end of ATC and the auricle of the right atrium (RAT) [17]. This procedure was first described by H. Wenden et al. in 1984 [17]. [17]. A study was conducted, which analyzed scientific publications in the PubMed database; our clinical material was compared with other materials, which provide the best evidence to answer the question.

Patient M., 45 years old, was admitted to the cardiosurgery department of the ASMI clinic on 12.01.22. for congenital heart disease, partial anomalous drainage of pulmonary veins with complaints of dyspnea, palpitation during exercise, general fatigue. The defect was detected in childhood, but the patient's parents refused surgery. Her condition worsened recently, when she noted increased dyspnea and palpitations. On admission to the hospital: the patient's general condition was moderately severe, no acrocyanosis, the examination showed capillary blood SaO2 of 90-92%. Clinical manifestation of the defect depends on the volume of blood discharge, its duration,

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the degree of pulmonary hypertension and other complications. Subjective symptomatology of childhood malformation is poor. Patients usually complain of dyspnea on physical load and increased fatigue. In adults, there are often signs of right ventricular insufficiency. The natural course of the defect is the same as in ASD. Auscultation revealed a medium-intensity systolic murmur in 2-3 intercostals on the left side of the sternum; the 2nd tone over the pulmonary artery was accentuated. The liver and spleen were not enlarged. According to the publication, PAPVD reveals a systolic murmur with its maximum intensity in the second-third intercostal space on the left side of the sternum. With large blood discharges, some patients have a gentle diastolic murmur over the tricuspid valve; II tone over the pulmonary artery is widely split irrespective of respiratory phases. In the presence of pulmonary hypertension, the pulmonary component of II tone is amplified. The electrocardiographic picture of PAPVD is nonspecific and reflects hypertrophy of the right atrium and ventricle, as well as overload of the latter. The degree of these changes depends on the amount of blood loss and the duration of the defect [8,15]. In our case the electrocardiogram showed sinus rhythm, electrical axis of the heart was deviated to the right, hypertrophy of the right heart, incomplete blockade of the right Giss bundle. Echocardiography noted: - normal heart position, small (20 mm) upper secondary septal defect with left-right shunt was detected. right heart dilatation, right ventricular wall thickening, small turbulent flow in the right atrium was detected. LP size. - 28 mm, LV end-diastolic dimension (EDD) - 34 mm, LV end-diastolic volume (EDV) - 54ml, LV end-systolic dimension (ESD) - 25 mm. LV ejection fraction (EF) - 58%, RV EF - 62%. Free RV wall thickness - 15 mm. The pericardial cavity was free. The insignificant disturbances of hemodynamics and clinical course of malformation with ASD make correct diagnosis difficult. Of all traditional methods, radiological examination is the most informative, allowing to recognize abnormal drainage of pulmonary veins into hollow veins quite often [3,8,12,14].

Accurate diagnosis of all forms of malformation is possible with correct and complete cardiac probing and angio-cardiography. It is necessary to differentiate the defect first of all with ASD and other malformations with increased pulmonary blood flow. Routine radiological examination reveals signs of increased arterial vascular pattern in the lungs and enlarged right heart and pulmonary artery chambers of varying severity. In addition to revealing general signs of malformation, in some cases it is possible to determine the type of abnormal pulmonary vein drainage. In cases of abnormal drainage into the superior vena cava, a radiograph in anteroposterior projection reveals a dilated shadow of the lower segment of the superior vena cava and enlargement of the right lung root. When the pulmonary veins infiltrate into the nephrogenic vein, a rounded shadow, inseparable in multiaxial view from the right contour of the vascular bundle, is revealed above the upper edge of the lung root [6,16]. In our case, X-ray examination of the chest organs revealed: - moderate arterial hypervolemia of the small circulatory circle (SCC); the heart was enlarged in cross section more to the right, the size of PP and RV was increased, the LA arch was bulging, the right atriovasal angle was shifted up, the retrocardiac space was narrowed at the atrium level in the 1st c/o. Cardiothoracic index (CTI) was 56.5%. In the preoperative period the patient had no abnormalities in the general blood count, there were normal values of urea, creatinine, glucose, potassium, sodium, total bilirubin, albumin, total protein, "C" reactive protein, AST, ALT, coagulogram.

The patient was prepared for surgery with the diagnosis of partial anomalous pulmonary vein drainage in the VPH on 18.01.22. The course of the operation. (operator - Candidate of Medical Sciences M.A. Mamatov) - correction of malformation under conditions of artificial circulation (AC), hypothermia (32°C) and combined pharmaco-hypothermic cardioplegia. Midline sternotomy; the pericardium was opened; the heart was enlarged at the expense of the right sections; the superior vena cava was dilated to 2.5 cm; the right pulmonary veins were drained into the ERV, the LV was isolated acutely and bluntly; the aortic diameter 2.5 cm, cannulation of the aorta and vena cava (the ERV was cannulated with "L" shaped cannula); IR apparatus (AIC) was connected, under hypothermic (32g) artificial blood circulation (73min), the aorta was clamped, antegrade pharmacocold cardioplegia was performed, for topical myocardial hypothermia snow "crumbs" were used, pharmacocold cardioplegia (58min), right atrium was opened, incision was extended to ERV, where the right LV was abnormally drained; the upper ASD was seen 2x1 cm, the latter dilated to 2x3 cm; a wide autopericardial patch was taken, the ASD was sutured so that the mouths of the right pulmonary veins drained into the LP; a leaflet patch was applied to dilate the ASD mouth; the incision of the PP was sutured with prolene 4. 0. . Hemostasis; Prevention of air embolism. Routine AIC arrest (101min). Drainage of mediastinum and pericardial cavity. Layered suturing of the wound.

The patient was transferred to the intensive care unit with stable hemodynamic parameters using inotropic support with dopamine (3 μg/kg/min) and dobutamine (0.3 μg/kg/min). The duration of artificial

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lung ventilation (ALV) was 4 h 30 min; dobutamine infusion was 8 h and dopamine infusion was 6 h. The postoperative period was smooth. The patient was discharged home on the 9th day after surgery in a satisfactory condition. According to pulse oximeter data, blood oxygen saturation was 96-99%.

RESULTS OF THE STUDY . At follow-up examination 3 months after surgery the condition was quite satisfactory, he had no complaints. Capillary blood SaO2 is 96-98%. ECG showed sinus rhythm. Normal position of EOS. Hypertrophy of the RV was decreasing. Radiological examination of the chest organs showed: pulmonary pattern is closer to normal, curve of PP and LA decreases. Left ventricular arch is almost normal. Cardiothoracic index (CTI) was 48.2%. Postoperative EchoCG determined the airtightness of MPP, no stasis in LV, no stenosis of ERV was detected. LV size - 29 mm. LV MVP increased from 34 to 39 mm; MVP - 60ml; LV CIR - 26 mm. LV FV 60%, RV FV 58%. The thickness of RV wall decreased from 15 to 13 mm. The tricuspid valve function was normal. The clinical status of the patient corresponds to the I class according to NYHA. The results of surgical correction of PAPVD of the right pulmonary veins in VPV demonstrate good immediate results and low mortality rate. In the long-term period the correction results are also good, the occurrence of such complications as sinus node dysfunction, ERV stenosis [12,16] is rare. In the early postoperative period, sinus node dysfunction after biplane technique was observed in 0-55% of cases, after Warden operation - in 0-7.1%. In the long-term follow-up period sinus node dysfunction persisted after biplane technique in 0-5% of patients, after Warden operation only in 1 patient 6 years later atrial flutter developed. Moderate atrial septal stenosis not requiring correction was observed in 0-8% of patients with biplane technique [9,11,15].

CONCLUSION. The risk of complications was the main motivation for the introduction of new surgical techniques. However, as we can see from the above reviews, any technique carries a potential risk of one or another adverse effect. The considered option of partial abnormal drainage correction in the EPO shows satisfactory results. The risk of developing EEPV stenosis is rarely encountered after biplate correction.The risk of VPV obstruction exists in any correction technique and dictates the need for caudal extension of the VPV. Pulmonary vein obstruction is a relatively rare event after PAPVD correction in the VEP; as a rule, it is asymptomatic, a diagnostic finding and does not require correction.

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