

TOTAL ABNORMAL PULMONARY VEIN DRAINAGE

(Literature Review)

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ANNOTATION

To the article " Total anomalous pulmonary venous drainage". In this scientific article, the authors describe a literature review of total anomalous pulmonary venous drainage (TAPD). In this regard, the study of the clinical course and diagnostic methods of TADLV is relevant, which makes it possible to timely detect the defect, rationally plan the examination and choose the optimal method of correction. The definitions, frequency of occurrence, causes of the defect and diagnostic methods are described, including echocardiography, multislice computed tomography with contrast and catheterization of the heart cavities with angiocardiology. Various types of surgery are described for various variants of TADLV . 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 . Literature review describes the complications of the defect and the results of the operation.

Keywords: congenital heart disease, diagnosis, total anomalous pulmonary venous drainage, atrial septal defect, vena cava, echocardiography, catheterization of the heart cavities with angiography, result .

DEFINITION

Total anomalous pulmonary venous drainage (TAPVD) is a complex congenital heart disease in which all pulmonary veins (PV) flow into the right atrium or its tributaries [7,35,38,41,46]. At the same time , communication between the large and small circles of blood circulation occurs at the level of the atria through the ASD. The first description of this vice belongs to J. Wilson [30].

FREQUENCY

Among all CHD , total ADLV occurs in no more than 0.7-2.6% of cases [10,20,29,36,45]. In adolescents and older children, the proportion of TADLV begins to decrease sharply, and in adults this defect occurs in isolated cases [20,33,39]. Although, McManus V.M. et al . [21], P . Arciprete et al . _ [5] observed and operated on patients in the 3rd or 4th decade of life. However, according to Amosov N.M. et al . [1], Andoh . et al . [6] the vast majority (80%) of infants die before the age of one year. Such a "malignant" course of TALVD is associated with early progressive heart failure, which occurs due to the small size of the ASD, stagnation of the ICC, and severe hypoxia.

CLASSIFICATION

Among the numerous classifications of ADLV, the classification proposed by R has received the greatest distribution . Darling et al . [12], which is based on the anatomical principle depending

on the level of the inflow of the PV. The authors distinguish 4 types of defect: Type I - with supracardiac or supracardiac : PV flow directly into the superior vena cava (SVC) or its tributaries; Type II - cardiac or cardiac: PV flow into the right ventricle or into the coronary sinus; Type III - infracardiac or subcardiac: PVs flow into the inferior vena cava (IVC) or its tributaries below the level of the diaphragm, and type IY - mixed : blood from the PV enters the right heart in two or three ways described above. Such differences in the anatomical structure of the defect determine the specifics of hemodynamic disorders, clinical course, tasks and methods of surgical treatment of each type of defect.

HEMODYNAMICS

Hemodynamics in total ADLV is characterized by volume overload of the pulmonary circulation (MCC) and depletion of the systemic circulation. In 2/3 of patients, pulmonary hypertension is observed [7,15,26], accompanied by cyanosis and early development of severe decompensation. Pulmonary arterial hypertension in 20% of patients is hyperkinetic in nature and sclerotic in 80%. When assessing hemodynamics, the presence of a pressure gradient between the right and left atria is of great importance , which indicates the inadequacy of the interatrial defect, the size of which determines the volume of systemic blood flow [23,28]. The more blood enters the ICC and the pronounced pulmonary venous obstruction, the faster the volume overload of the right heart and arterial PH develop [6,4 0]. At the same time, the left parts of the heart retain their usual size or are hypoplastic . The stroke volume of the left ventricle and the minute volume of the systemic circulation (BCC) often decrease [2,8,16]. Often, the TALV clinic is characterized by the development of early and rapidly progressive heart failure , which is the main cause of high mortality in patients, many of whom die before the age of one [4,1 4,19]. The general condition of patients is severe, and only in isolated cases - satisfactory [3,13,40] . Modern research methods are used in clinical diagnostics : electrocardiography [4,9,25,46]; echocardiography [9,17,18,33,34]; radiography [31,38,39,43,44,46]; catheterization of the heart cavities with angiocardiology [25,30,33,34,40,44]. There is evidence in the literature that in the diagnosis of ADLV, cardiofibroscopy , MSCT and magnetic resonance imaging were used [6,16]. This method provides quite a lot of information about the presence of an ASD and abnormal pulmonary venous drainage.

NATURAL FLOW AND FORECAST

In the natural course of TALV with PH, the mortality rate is very high. The average life expectancy of these patients is only 7 weeks; moreover, the presence of an obstruction in the outflow tract from the PV reduces the survival rate to 3 weeks [6,2 3,44]. According to many authors: R . Ashoush et al . [7], J . Davis et al . [13], at least 75% of patients die in the first year of life, and only a few patients survive to 3-4 years of age. In contrast, the natural course of TALV in patients with normal LA pressure is relatively favorable; the disease proceeds, as with a large ASD. There are reports in the literature describing patients who survived to 50–60 years [21].

TREATMENT

With TALV, radical surgical treatment of the defect is actually the only intervention after which a stable positive result can be expected. Nevertheless, palliative operations do not lose their significance [25,34,40], although they can only be used to treat newborns in critical condition - the operation consists in increasing the interatrial communication by balloon arterioseptostomy, which results in better conditions for blood supply. vi in the LP, and consequently, the improvement of blood circulation in the systemic circle [30,33,44]. However, the operation has limited efficacy and is performed in children under three months of age. In recent years, other types of palliative interventions have been introduced into clinical practice, which can be used in the narrowing of the common collector ADLV: the essence of the method is endovascular balloon dilatation of the narrowed area of the common collector [25,33,44,45].

The general principles of radical correction of TALVV are reduced to the creation of a wide anastomosis of the pulmonary veins with the LA, closure of the ASD and ligation of the PV collector [3,4,5,7,8,20,35,39]. The methods of surgical treatment of the defect are different depending on the anatomical form of TADLV. Radical correction is usually performed under EC conditions, and in newborns, deep hypothermia with circulatory arrest is also used [4,8,20,37,42]. There are two options for the operation. The first option is indicated for severe LV hypoplasia: in order to prevent postoperative heart failure, ASD is partially eliminated by using a perforated patch. It is possible to use a two-stage tactic: the first stage is anastomosis between the common collector of the PV and the LA; after the development of the left sections, the next second stage is the ligation of the common collector at the place where it flows into the systemic vein. The second option is the simultaneous correction of all components of the defect under EC conditions of TADLV I and III by R. Darling' et al. [12], which are characterized by the presence of a common LP collector, perform similar operations. The operation is usually performed from a median sternotomy, under EC conditions. Anastomosis between the LA and the PV collector can be performed by accessing the heart through the RA and extracardiac [11]. The area of the anastomosis should not be less than the atrioventricular orifice [3,5,7,38,43], because small sizes from the mouth can lead to stagnation of blood in the PV; after anastomosis, the ASD is closed with a patch of autopericardium or synthetic tissue; Moreover, with a small size of the RA cavity, the cavity can be enlarged by moving the lower edge of the patch towards the RA [8,20,37,40,46]. Later, the operation was modified by N.M. Amosov et al. [1], B. Barrat - Boues - et al., [8]. Rear access to the LP, proposed in 1970 by V. Roe, did not receive recognition.

Correction of the cardiac type of defect according to the method J. Kirklin with et al., [18] is the simplest in execution technique [20]. In case of confluence of the common pulmonary vein collector into the RA, its orifice is usually wide, the correction consists only in moving the orifice of the PV into the LA with simultaneous plastic surgery of the ASD. When the ADLV flows into the CS, a similar operation is applied with the VanPraagh modification with et al. [27] - first, a part of the septum between the oval window and the KS is excised, then the anterior wall is dissected, as a result of which the newly created wide ostium of the KS is mixed into the LA; the patch is sutured in such a way that, after fixation, its orifice of the CL moves into the cavity of the LA [28,38,44]. A frequent postoperative complication with this type of correction is conduction disturbance with various forms of brady rhythm, which is apparently associated with damage to the intra-atrial internodal pathways during resection of the atrial septum. In

order to prevent this complication, in 1972, VanPraag et al . proposed a technique consisting in resection of the "roof" of the CS followed by suturing the orifice of the CS and ASD [28,38,44]. However, Reed G . E . _ et al . , [22], using this surgical technique, did not note cases of a decrease in bradycardia in the postoperative period. At the same time, another serious complication of this modification was revealed - PV stenosis in the area of the collector or the orifice of the CL, which often develops 3-4 months after the operation. Thus, the frequency of PV obstruction in TALV in the CS after defect correction , according to C . M . _ Whight with et al . , [31] occurred in 3.6% of cases . K. Turley s et al . , [26] observed obstruction of the pulmonary veins in 60% | observations, aJ . Davis et al . , [14] - in 10% and G . Reed et al . , [22] - in 22% of cases.

Particular technical difficulties usually arise when correcting the infracardial and supracardial forms with abnormal drainage directly into one of the vena cava: a complicating point in such cases is that the PV and LA are far apart and therefore it is necessary to perform complex reconstructive operations, which were recommended by a number of authors [1 8,26,46] . At the same time, a tunnel is created in the SVC or IVC with the help of a patch, or by longitudinal division of its cavity, carrying arterial blood; in this way, the formed channel is inter- atrially mixed through the ASD into the LA cavity [4,7,46].

With a mixed type of defect, the method of corrective surgery is more diverse. Depending on the specific anatomical forms, it consists of a complex of operations performed with total and partial ADLV. The success of all types of operations depends on the creation of an adequate - anastomosis between the LA and the pulmonary veins, on careful prevention of air and arterial embolism [7,2 0,43,44].

Mortality after surgical correction of TALV, according to a number of authors: Friesen C . L . _ H . , et al . [44], C . M . _ Whight with et al . [31], S . Willi ams et al . [29] ranged from 25 to 84%, and has recently decreased to 6% [7,8,45]. Significantly lower mortality can be achieved in children under 3 years of age [4]. Lethal outcomes are usually associated with the initial serious condition of patients operated on against the background of severe PH and arterial hypoxemia [29,43,46]. Mortality after surgery in children under 6 years of age according to M. Gomes with et al . [17] was 10%. It is higher in the infracardial form of abnormal pulmonary venous drainage in the IVC, in PH, and hypoplastic left heart.

LONG-TERM RESULTS

And the study of long-term results is one of the most reliable indicators of the feasibility of a particular treatment method, which allows you to evaluate the technique from the standpoint of time. Long-term results of surgical treatment of ADLV in the literature are covered in single works and more often on a small material [3 3,44,45].

One of the first, M. Gomes et al . [17] paid close attention to the results of surgical correction in the long-term period after TALV. They examined 49 patients, out of 59 operated, in the period from 1 to 14 years. One patient died 2 years after surgery from arrhythmia. Four in the long-term period underwent an ACG study with suspicion of a residual shunt, and in two of them it was detected (6 and 14 years after the operation). Systolic pressure in the pulmonary artery in all examined patients was within 30 mm Hg .

According to A. Serra et al . [23] and A . Serraf with et al . [24] , PV stenosis is a serious complication for patients before and after radical correction of TALV. The authors present the

results of surgery in 107 patients, 28 of them with subcardial and 23 with supracardial forms, of which 57 had collector stenosis. In the postoperative period, 20 patients died (18%). In the long term, obstruction at the site of the anastomosis was found in 4 patients [3 2]. One patient died without surgery out of two reoperated, one also died in the postoperative period. In the latter case, the patient underwent balloon dilatation of the obstructive area. A similar case of successful transluminal balloon dilatation of the stenotic area of the collector in patients after surgery was described by other authors [3 2,43,46].

Extensive experience in the treatment of TALV is described by R. Lambetal .[19] - 80 patients. In the remote period, 6 patients died . The cause of five deaths was obstruction of the venous collector (5 weeks to 3 months after surgery). In two cases, stenosis of the CS opening was noted (1 with mixed, the second with ADLV in the CS); in two cases due to stenosis of the PV collector (with supracardial and subcardial forms) and in one case there was a stenosis of the opening of the right PV, which led to edema of the right lung (with cardiac form in RA). The pressure in the LA in these patients was from 65 to 120 mm Hg. Art. The cause of death of the sixth patient was the obstruction of the SVC with TALV into the superior vena cava. Reoperations were performed in 8 patients (three with obstruction, 4 with ASD recanalization, and one with SVC obstruction). One of the conclusions made by the authors is that the correction technique for TADLV in the knee joint, proposed by R. VanPraaghetal . [27] is controversial.

FriesenC . L. _ H. _ etal . [44] describe long-term results in 28 patients up to 14 years. Five patients had stenosis of the venous collector, which required repeated surgical intervention (in three cases, the operations ended in death). Another reason for reoperations in 4 patients was ASD recanalization . How do you tell Y . Andoet a1. [6], in the long-term period, out of 38 neonates operated on with supracardial TALV according to the Gerson y method, there were three cases of narrowing of the anastomotic site, two of which ended in the death of the patient. They used the so-called "double-patch" technique on the ASD and, of course, a patch that expands the LA. There were no deaths in the postoperative period and all patients felt well.

There are specific complications associated with impaired blood flow through the SVC due to various reasons - narrowing of its lumen after division, wrinkling of patches, thrombosis. HusainS . A. _ etal . [45] noted signs of temporary blockage of the SVC in one patient . C. _ Chartrandetal . [10] stenosis of the SVC was found in one patient with an accessory SVC 5 years after ADLV correction. V.S. Sergievsky et al . [2] reported a case of thrombosis leading to cerebral edema. SVC blood flow disorders can end safely and be compatible with life [10,45], despite narrowing or even complete obstruction [4,1-5]. The blood flow in this case is carried out along the collaterals through the system of the unpaired vein in the IVC or through the additional SVC, if any.

Based on the foregoing, it becomes clear that there are many unresolved problems in the treatment of abnormal pulmonary venous drainage, among which are the risk of narrowing of the pulmonary-atrial anastomosis, SVC, impaired pulmonary venous outflow, preservation of the left-to-right shunt, and cardiac arrhythmias.

Thus, total anomalous pulmonary venous drainage is a rare and complex heart disease with a fairly typical clinical presentation and early development of pulmonary hypertension. In the timely diagnosis of the defect, the leading place belongs to echocardiography, radiography, MSCT and catheterization of the heart cavities with ACG. Early and adequately performed

reconstructive intervention with dislocation of the pulmonary veins to the left atrium allows to obtain good long-term results.

LITERATURE

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