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MODERN SURGICAL CORRECTION OF PULMONARY ARTERY ATRESIA WITH MAJOR AORTO-PULMONARY COLLATERAL ARTERIES AND INTERVENTRICULAR DEFECT IN CHILDREN

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Abstract*: A case of successful treatment of a six-year-old girl with a complex congenital heart disease, rare at this age. The observation shows that atresia of the lungs with a defect in the ventricular septum and large aortopulmonary arteries is a complex group of diseases and requires phased unifocalization and complete recovery in infancy. Since the blood flow to the lungs is completely dependent on these arteries, at a later age in children with the natural course of this complex heart disease, the results are unpredictable. However, since this heart disease is associated with more than 60% mortality by 10 years of age, the authors proposed a change in approach: unifocalization of the pulmonary artery into the right ventricle as the first stage of the procedure. Thus, avoiding complications, timely determining adequate treatment tactics, improving the quality of life.*

Keywords: *major aorto-pulmonary collateral arteries, unifocalization of pulmonary blood flow, atresia of the pulmonary arteries.*

INTRODUCTION: Pulmonary atresia (PA) is a congenital absence of direct communication between the right ventricle (RV) and the pulmonary artery system (PA). This is a rather rare CHD, which is observed in 2-3% of all CHD cases. The defect occurs both in the form of a combination with an interventricular septal defect (VSD), and with various complex CHD, such as transposition of great arteries, atresia of the right atrioventricular opening, a single ventricle of the heart, etc. The anatomical criteria of the defect are as follows: 1) the absence of PA at various levels; 2) large VSD; 3) the presence of additional sources of collateral blood flow lungs; 4) hypertrophy of the pancreas; 5) dextroposition of the aortic root; 6) normal relationship between the aorta and the LA trunk [1, 2,6,8,19,24,30].

ALA with VSD is a congenital heart disease, and the child's condition mainly depends on the size of the patent ductus arteriosus (PDA), the presence of large aortic-pulmonary collateral arteries. Hemodynamic disturbances in this defect are mainly determined by the lack of direct communication between the RV and the PA system, while the direct flow of venous blood from the RV is impossible. The blood flow of the lungs is carried out by a roundabout way from the pancreas through the VSD into the left ventricle (LV), and then the mixed arterialized blood enters the aorta and only then through the PDA or through the collateral vessels into the lungs. At the same time, blood oxygen saturation in the aorta, collateral arteries and pulmonary artery is identical [4, 5, 7, 11, 13, 14, 16]. The prognosis of a patient's life depends on the nature of pulmonary blood flow. The mortality rate of children with ductus-dependent hemodynamics up to 12 months is 90%. In patients with several sources of pulmonary blood flow and moderate cyanosis by 3-5 years of age, the mortality rate is 50%. With increased pulmonary blood flow and the presence of large aorto-pulmonary collateral arteries, patients die when pulmonary hypertension develops, mainly in the third decade of life. In general, the median survival rate for patients with ALA and VSD is within 6 months. - 2 years.

During the staged treatment, sequential unifocalization of pulmonary blood flow for each lung is performed with further reconstruction of the outflow path from the right ventricle and closure of the interventricular defect [1, 2, 8, 15, 16, 17, 21, and 22]. Simultaneous radical intervention allows you to eliminate the multifocal nature of the blood supply to the lungs and perform intracardiac correction immediately [6], which, according to VM Reddy et al. [9, 23, 25, 26, 29], possibly in 90% of patients. The difficulty in choosing the optimal treatment tactics for patients with this pathology can lead to unreasonably high mortality [21, 22, and 30].

Case report: Here is a case from our practice with a radical correction: Patient K.Kh. GN ID 12953427 (Artemis hospitals, Gurugram, India) DOB October 2013, On December 16th 2019 was admitted to our hospital with a diagnosis of Atresia of the pulmonary artery. After comprehensive examination, following diagnosis was established: "Congenital blue heart disease. Tetralogy of Fallot, Pulmonary atresia. Narrowing of the right pulmonary artery. Open patent ductus arteriosus (PDA). Major aorto-pulmonary collateral arteries (MAPCA). Normally functioning left ventricle" Prepared for the surgery. The general condition is severe, afibril, vital parameters are within normal limits, SpO_2 saturation in room air is 64%.

The surgery was performed under general anesthesia and cardiopulmonary bypass. After processing the surgical field, a median sternotomy was performed. After median sternotomy, in order to improve exposure, the thymus was removed. Pericardium was opened and fixed using 2-0 temporary silk sutures. In the course of the manipulation, adequate hemostasis was performed. Severe Right ventricular hypertrophy (RVH), Pulmonary atresia, very small segment MPA, Confluent branch pulmonary arteries, Small RPA (~3mm), adequate size of LPA were noted. Posterior mediastinum was entered through the space between the SVC and ascending Aorta. Using blunt and sharp dissection two large (4 mm) essential MAPCA`s going to the right lung were identified and looped with silastic tapes. One non-essential MAPCA to the left lung was also identified and closed with a medium ligature clip. Both MAPCA`s to the right lung were dissected in their entire length.

Cardiopulmonary bypass (CBP) was established by catheterization of the SVC, IVC and aorta using 5-0 prolene purse string sutures, all cannulas were deaerated and connected to the corresponding ends of the CBP circuit. A cardioplegic needle was inserted into the ascending part of the aorta, and was also deaerated and connected to the suction casing as an aperture of the aortic root. IR was created with good returns and close full flow rates.

Ascending aorta, MPA, RPA and LPA were mobilized on CPB. Both Right lung MAPC`s were closed at their origin with medium ligature clip and divided using sharp scissors. Stumps were re-enforced with 6-0 prolene sutures. MAPCA from the distal arch/proximal descending aorta was long and was well suited to the superior surface of RPA. MAPCA from the mid-descending thoracic aorta was short, running under the carina and after mobilization, reached the posterior surface of RPA. A long arteriotomy (extending from the hilum to the RPA/LPA) confluence was performed on the superior surface of RPA and splayed open using 7-0 prolene stay sutures.

Working through this incision a 6 mm arteriotomy was performed on the posterior surface of RPA and the MAPCA from the descending thoracic aorta was anastomosed to this posterior arteriotomy on 7-0 prolene sutures -end to side fashion.

The superior MAPCA was anastomosed to the superior surface of RPA in its entere length (side to side) using 7-0 prolene sutures – enlarging the RPA from its origin to the hilum.

LPA was dissected out and between stay sutures -a longitudinal arteriotomy was performed on

the small MPA segment and this was extended on the LPA.

A 100 PTFE conduit was cut to an appropriate size and has been used as RV to PA conduit. This conduit was sutured to an incision on the LPA using 6-0 prolene sutures. SVC, IVC were snared, aortic root was vented and a sharp head down position a right ventriculotomy was performed about 2-3 cm to the right of the LAD. The RV to PA conduit was sutured to this ventriculotomy using 6-0 prolene sutures. Patient was rewarmed to 36 ºC and on inotropic support, she was easily weaned off bypass. About 500 cc of ultra-filtrate was removed using "Modified ultrafiltration". Following reversal of heparin by protamine, all cannulas were removed. Purse strings were tied down and reinforced with 5-0 sutured. To prevent gastric fibrillation, two wires of a pacemaker were left on the right atrium and on the right ventricle. The wounds were sutured in layers 3-0 with vicryl.

The duration of the surgery was 7 hours, CBP time 218 min. The wound healed by first intention; the child was discharged home on the 7th day after the surgery, under outpatient supervision.

Resume: Pulmonary atresia with VSD and MAPCAS is a complex subset of disease and needs staged unifocalization and complete repair in infancy.

However, blood flow to the lungs is completely dependent on these MAPCAs that narrow down with time due to medial hypertrophy or increase the pulmonary vascular resistance of the part of the lungs that they supply and so, in late presenting children with this heart condition - the outcomes are unpredictable.

Since this heart condition is associated with more than 60% mortality by 10 years of age- we have modified our approach and we offer unifocalization as a stage one procedure with a right ventricle to pulmonary artery conduit.

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