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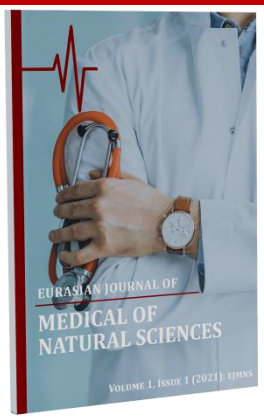

EARLY NEONATAL, POSTNATAL DIAGNOSIS, CLINICAL MANIFESTATION, TREATMENT AND PROGNOSIS FOR TETRAD FALLOT

Xadjimukhamedova Z.B.
Alieva N.R.
National Children's Medical Center
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ABSTRACT

The article presents literature data on the frequency, main clinical manifestations, modern methods of early neonatal and postnatal diagnosis, treatment and prognosis of tetrad Fallot is a congenital defect of the cardiovascular system.

Tetrad of Fallot is a complex anatomical anomaly of the heart, which in typical cases includes 4 components: pulmonary artery stenosis, ventricular septal defect (LVD), dextroposition of the aorta and hypertrophy of the right ventricle. According to the International Classification of Diseases of the 10th revision, there are: Q 21.3 Tetrad of Fallot - a defect of the interventricular septum with stenosis or atresia of the pulmonary artery, dextroposition of the aorta and hypertrophy of the right the ventricle. The first reports of the defect belong to M. Stensen (1673). A.A. Kisel (1887) was the first to carry out a lifetime diagnosis of the defect. French Doctor Etienne-Louis Arthur Fallot (1888) coined the term "tetralogy" to clinically designate the four components of the "blue" disease. Tetrad of Fallot is the most common heart defect of the "blue" type (75%), its frequency is: among all congenital heart defects — 7-13% according to clinical data, 15-16.7% according to pathological data, in newborns — 5-8%, in high school age — 12-14%. The tetrad of Fallot is found with numerous syndromes with any type of inheritance (autosomal dominant, autosomal recessive, X-linked recessive).

The clinic of the tetrad of Fallot may be different, due to the variability of hemodynamic disorders. The severity of hemodynamic disorders and the severity of the course of the defect are primarily determined by the degree of narrowing of the pulmonary artery, which can range from minor stenosis to its complete atresia. Most often, with this defect, pulmonary artery stenosis is infundibular (low, high, or in the form of diffuse hypoplasia) or combined (a combination of infundibular stenosis with valvular stenosis, hypoplasia of the ring and trunk of the pulmonary artery). Atresia pulmonary artery valves, an extreme form of obstruction, is much less common. Anatomical obstruction of the excretory part of the right ventricle may be combined with hypoplasia and stenosis of the branches of the pulmonary artery, possibly with atresia of the left branch of the pulmonary artery. The tetrad of Fallot is characterized by a high large defect of the interventricular septum (perimembranous subaortic) and dextroposition of the aorta, that is, the displacement of the mouth of the aorta to the right so that it "sits astride" the interventricular septum, and from the right ventricle there is a direct

exit into the aortic lumen. So Thus, two blood streams enter the aorta — from the right ventricle (venous) and the left ventricle (arterial).

The fourth sign of the defect is hypertrophy of the right ventricle, which is a secondary compensatory component. The development of right ventricular hypertrophy is especially influenced by its adaptation to pressure in the aorta. With severe pulmonary artery stenosis, blood flow is low. A significant part of the venous blood from the right ventricle is discharged into the aorta, causing the appearance of general cyanosis in the child. The discharge from right to left (venous-arterial) increases in a child with physical exertion, since blood flow to the heart increases significantly at this time, and blood flow through the lungs it can practically not increase due to pulmonary artery stenosis. As a result, all excess blood entering the right ventricle is discharged into the aorta, increasing cyanosis. During physical activity, the oxygen saturation of the blood in the aorta can decrease by up to 60%. At rest , the blood flow through the lungs can almost match the inflow, being sufficient. In this case, venous blood is almost not discharged into the aorta, while the oxygen saturation of the blood in the large circulatory circle remains high. With moderate pulmonary artery stenosis , other hemodynamic disorders are noted. At rest, on the contrary, blood is discharged from left to right (arteriovenous) through DMJP — from the left ventricle to the right, in this case the pulmonary blood flow will be increased. During physical activity, blood flow to the heart increases significantly, but pulmonary blood flow as a result of pulmonary artery stenosis remains the same as at rest. Excessive the amount of venous blood will be discharged from the right ventricle into the aorta (venous arterial discharge). Two opposite mechanisms of hemodynamic disorders — from the discharge of blood from right to left in the "blue" forms of the defect before the discharge of blood from left to right with "white", lighter versions of the tetrad Fallot — cause a variety of different clinical manifestations of this defect. Manifestations of extra-cardiac compensation of the defect include the development of polycythemia, while the number of red blood cells can reach up to 8 T / l, the hemoglobin content increases to 250 g / l. Gradually compensatory blood circulation develops between the large circle and the lungs, carried out mainly through the dilated arteries of the bronchi, chest, pleura, pericardium, esophagus and diaphragm

Early neonatal and postnatal diagnosis Severe course of the defect with cyanosis in the neonatal period is observed in 1/3 of patients. Cyanosis appears more often from the second half of life. Dyspnea-cyanotic seizures usually occur in the 2nd, 3rd years of life and are accompanied by pronounced weakness, deep rapid breathing, loss of consciousness. The reason for the appearance of such attacks is the temporary closure of the outflow pathways from the right ventricle — a spasm of its infundibular section, as a result of which all venous blood enters the ventricle through the DMF it causes or enhances hypoxia of the central nervous system. At the same time, the intensity of systolic noise along the left edge of the sternum decreases. It is noted that the weaker the noise, the more severe the anatomical variant of the defect. In rare cases (4-5% of cases) , systolic systolic noise can be heard on the back between the shoulder blades due to the development of collateral circulation between the vessels of the large and small circulatory circles. The second tone on the pulmonary artery is weakened. The depletion is determined radiologically pulmonary pattern. The shadow of the heart is small in the shape of a boot, with a depression in the area of the arch of the

pulmonary artery. An electrocardiogram (ECG) reveals a significant deviation of the electrical axis of the heart to the right, signs of hypertrophy of the right ventricle and right atrium (an increase in the amplitude of the P wave), a slowdown in the period of atrioventricular conduction. On phonocardiography (FKG), intense systolic noise is recorded, the second tone above the pulmonary artery is widely split, and the degree of splitting depends on the degree of stenosis of the pulmonary artery, pulmonary the component of the second tone is weakened. With pulmonary artery atresia, noise may not be detected.

During ultrasound examination, the following changes are characteristic: — when scanning along the long axis of the heart , a violation of the continuation of the interventricular septum into the anterior wall of the aorta is determined; — dilation of the aorta (Fig. 1) and its location closer to the anterior chest wall, as a result of displacement, the aorta has the appearance of a "rider" or "sitting astride" aorta; — the presence of a large ventricular septal defect; — myocardial hypertrophy and increased trabecularity of the right ventricle; — stenosis of the right ventricular outlet tract and pulmonary artery obstruction; — interruption of the ultrasound beam during the transition from the cavity of the left ventricle to the aortic root when M-scanning; — turbulent flow in the right ventricle during systole as a result of blood shunt from the left to the right ventricle; — turbulent flow in the outlet tract of the left ventricle into the diastole due to shunting of blood from the right to the left ventricle; — increased blood flow rate and turbulent flow in the pulmonary artery due to the presence of stenosis.

Criteria for postnatal diagnosis

Clinical:

— cyanosis, which can be observed from the first months of life, but more often appears by the year and later, increasing with physical exertion, emotional stress, crying, screaming, etc., skin color with cyanosis varies from blue to purple;

— the child squats down (typical of the tetrad of Fallot and rarely found with other "blue" vices) or lies with his legs brought to his stomach;

— low birth weight, developmental delay;

— dyspnea-cyanotic (hypoxemic) attacks, during which there is increased cyanosis, shortness of breath, tachycardia, anxiety, weakness develop, sometimes loss of consciousness. The duration of seizures ranges from a few minutes to 10-12 hours. Possible violation of cerebral circulation; — thickening and reshaping of nails in the form of "watch glasses" and nail phalanges ("drumsticks"), expansion of skin capillary networks on veins and in the forehead, epigastric pulsation;

— systolic tremor in the lower part of the chest on the left; there may be a heart hump;

— the boundaries of the heart are expanded not only to the right, but also moderately to the left;

— during auscultation , a rough systolic noise typical of pulmonary artery stenosis is determined, reaching a maximum in II–III intercostals on the left near the sternum (can be heard in the III–IV intercostals). The intensity of the noise varies and is inversely dependent on the degree of pulmonary artery stenosis. In children with significant obstruction of the right ventricular outlet tract, pronounced cyanosis and mild systolic murmur are detected, and in children with a lesser degree of pulmonary stenosis against the background of the

absence of cyanosis, a prolonged loud systolic is heard noise, sometimes accompanied by systolic tremor of the chest in the area of the right ventricular outlet tract;

— the second tone on the pulmonary artery is more often weakened or represented by an aortic component

— a one-component second tone (closure of the pulmonary artery valve is not heard).

Paraclinical:

— clinical blood test

— compensatory polycythemia due to high erythrocytosis (up to 8 T/l) and polyglobulia due to an increase in hemoglobin content (up to 250 g/l);

— radiologically: the shape of the heart in the form of a wooden shoe due to the rounded tip raised above the diaphragm and the occlusion of the arch of the pulmonary artery.

This shape of the heart is due to the hypertrophied right ventricle, which occupies the entire anterior surface and pushes the left ventricle posteriorly and upward. Hypertrophy of the right ventricle is detected in the lateral and second oblique projections. There is a slight increase in the size of the heart (since as a result of a decrease in pulmonary blood flow, congestive heart failure does not develop), dilation of the right atrium, right-sided aortic arch (in 25% of patients), increased transparency of the pulmonary fields due to depletion of the pulmonary pattern. 1/3 of the children have there may not be obvious impoverishment at an older age , and even a slight increase and chaotic looping of the pulmonary pattern due to a pronounced collateral vascular network are detected;

— ECG: a typically pronounced deviation of the electrical axis of the heart to the right (alpha angle from +120° to +180°), signs of hypertrophy of the right ventricle, right atrium (elevation and sharpening of the P–wave in leads II and V1-2);

— FKG: the second tone above the pulmonary artery is reduced in amplitude and narrowed due to weakening of the pulmonal component. There is also a high-frequency, medium amplitude spindle-shaped systolic noise with a peak in the first half of the systole;

— echocardiography: the location and expansion of the base of the aorta and the cavity of the right ventricle, reduction in the size of the left ventricle and left atrium, hypertrophy of the right ventricle, interruption of echo signals in the interventricular space 1. Examination of the newborn's heart. Scanning along the long axis of the heart. The aorta (AO) is dilated, its location above the interventricular septum ("sitting astride the aorta"), LV is the left ventricle, LA is the left atrium, RV is the right ventricle. The defect of the interventricular septum is indicated by arrows

Differential diagnosis with tetrad of Fallot should be performed with transposition of the aorta and pulmonary artery, double divergence of the main vessels from the right ventricle, a single ventricle, a two-chamber heart with pulmonary artery stenosis, and a common arterial trunk. Treatment. To stop a dyspnea cyanotic attack, it is recommended to position the child with the knees brought to the chest to reduce the venous return of blood to the heart. Constant oxygen inhalation is carried out, cordiamine is subcutaneously injected at 0.02 ml / kg of weight and a 1% solution of promedol at 0.1 ml per year life subcutaneously or intramuscularly. To relieve pulmonary artery spasm, obsidan 0.1 mg / kg is prescribed intravenously drip at a rate of 0.005 mg/min. The use of cardiac glycosides is not recommended, as they increase the tendency of infundibular stenosis to spasm. If the

measures are ineffective, an emergency surgical intervention is performed — the imposition of a systemic pulmonary anastomosis (between the aorta and the pulmonary artery). Surgical treatment is indicated for all patients with Fallot's notebook. Radical correction of this early malformation provides stable long-term survival in 98% of patients and freedom from repeated surgery in 63% of patients within 10 years.

Forecast. The average life expectancy in non-operated patients is 10-12 years. Palliative surgery lengthens the life expectancy of patients, but does not improve the prognosis. After radical correction of the Fallot tetrad in the absence of postoperative complications, the prognosis is quite satisfactory.

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