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Pathogenesis, Incidence, And Pharmacotherapeutic And Surgical Aspects Of Tetralogy Of Fallot

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Abstract. Tetralogy of Fallot is a pathology associated with a Cyanotic congenital heart defect. Tetralogy of Fallot is characterized by hemodynamic disorders, reduced pulmonary blood flow and venous return from the right ventricle to the aorta. The main symptoms of the pathological condition are pallor of the skin, signs of cyanosis, heart murmurs, delayed regurgitation, respiratory and heart failure. This article discusses the etiology, epidemiology, clinic of Tetralogy of Fallot, as well as modern principles of pharmacological and cardiosurgical treatment of the disease.
Keywords: Tetralogy of Fallot, dispo-cyanotic attack, classic form, severe form, pale form, ventriculoseptal defect, ventricular outflow tract obstruction, aorta dextraposition, right ventricular hypertrophy

Tetralogy of Fallot is a congenital heart defect that ranks high among combined defects and is manifested by the following main features: a defect in the interventricular septum, pulmonary artery stenosis, right displacement of the aortic orifice (dextraposition), and right ventricular hypertrophy. Tetralogy of Fallot occurs as a result of anomalous, i.e. abnormal development of the septum of the arterial cone of the embryonic heart. This heart defect is clinically manifested by cyanosis, shortness of breath, and developmental delay that begin at birth. Cyanosis and shortness of breath may sometimes increase for no apparent reason, and the child may even lose consciousness. If the pulmonary trunk is less narrowed, cyanosis may not be present [1]. In cardiology, tetralogy of Fallot occurs with a frequency of 7-10% among all congenital heart defects and accounts for half of all cyanotic-type defects. A detailed anatomical description of this defect as an independent nosological form was first studied by the French pathologist and anatomist E.L.A. Fallot, who later named the defect after him [19].

Classification of disease severity:

a) Severe form, characterized by severe cyanosis and shortness of breath from the first days of life, often from 1 year of age;

b) The classic form - this is when the cyanotic child begins to walk and engage in active movement;

c) Severe form, combined with shortness of breath and cyanotic attacks, in which the disease can occur from 3 months of life;

d) The clinical significance of the pale form occurs without arterial hypoxemia.

Life expectancy with the "pale" (pale) form of the defect is slightly longer than with the cyanotic form. Usually, patients who have not undergone severe surgery die from cerebral thromboembolism with the formation of an abscess, heart failure, and the development of infective endocarditis [3,5]. Also, in pathological conditions, dyspnoea-cyanotic attacks occur. Clinical symptoms are characterized mainly by the development of hypoxic attacks in young children (from 6 months to 3-4 years). During an attack, most often in the morning, the child suddenly becomes restless, cries, assumes an orthopneic position, bends his legs to the abdomen,

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and lies on his side. At the same time, cyanosis and shortness of breath increase, and the systolic murmur of pulmonary artery stenosis disappears. The duration of a dyspnoea-cyanotic attack can last from several minutes to several hours. In severe cases, loss of consciousness, cerebrovascular disorders, convulsions, and even death are observed. The following compensatory mechanisms develop in response to hypoxemia resulting from a stroke:

- ✓ Brain filling with blood
- ✓ Significant collateral circulation in the lungs
- ✓ New blood vessel formation in the myocardium
- ✓ Increased hemoglobin and red blood cell count
- ✓ Increased blood clotting
- ✓ Metabolic acidosis occurs

In emergency therapy, the child is first sedated, his clothes are unbuttoned. Humidified oxygen inhalation is performed through a mask. From pharmacological drugs, a 1% solution of promedol is administered subcutaneously at a dose of 0.1 ml/year. Cordiamine is administered subcutaneously or intramuscularly at a dose of 0.02 ml/kg. If there is no effect, it is recommended to slowly inject a 0.1% solution of obsidian at a dose of 0.1-0.2 mg/kg in 10 ml of 20% glucose solution (at a rate of 1 ml/min or 0.005 mg/min) [20].

The presence of systemic pressure in the right ventricular cavity of the heart of a patient with tetralogy of Fallot over time leads to the formation of pathological hypertrophy of the right ventricle, including the development of fibrous changes in the ventricular wall. This reduces the functional capacity of the right ventricle, which is very important for patients with tetralogy of Fallot who have been treated for a long time. In the first weeks of life, patients with tetralogy of Fallot may experience increased cyanosis. This is due to an increase in the degree of obstruction of the right ventricular outflow tract due to hypertrophy of the right ventricular myocardium. Most patients with tetralogy of Fallot have mild hypoplasia of the pulmonary artery valve annulus fibrosis [7]. The number of cardiomyocytes doubles by the 21st day of life, and then their number does not change significantly [10]. It is known that there are different types of stem cells in the body. One of them is called native cardiac stem cells or progenitor cardiomyocyte cells. One of the subtypes of these cells is cardiac mesenchymal cells. The progenitor cardiomyocyte cell population has signs of cardiomyocyte differentiation, but they do not express the sarcomeric proteins characteristic of cardiomyocytes. This type of cell is currently being actively studied [8,11]. These cell models are being used to study the mechanisms leading to congenital heart defects and the ability of such cells to participate in cardiac regeneration and repair, especially in patients with congenital heart defects. It has been found that cells from newborn patients with various congenital heart defects have a significantly greater regenerative and proliferative potential than cells from older patients [13]. Other studies have shown that cells from young patients have a higher regenerative potential than cells from older patients, which makes early treatment of patients with congenital heart disease more effective in terms of tissue regeneration [2]. Tetralogy of Fallot is a symptom complex that occurs with the syndrome of pulmonary valve agenesis. As a result of severe insufficiency of the projection of the fibrous ring of the pulmonary artery valve, a significant expansion of the central pulmonary vascular bed occurs, which leads to compression of nearby structures, i.e., the trachea, bronchi by the main and lobar pulmonary vessels [16]. For this pathology, it is typical that pulmonary syndromes prevail over cardiac syndromes. The syndrome of the absence of the pulmonary artery valve is characterized by a severe clinical course. Currently, there are several surgical approaches to correct this defect. The main goal of these methods is to correct intracardiac changes and eliminate pressure from the pulmonary arteries on the mediastinal organs. One of the most promising methods is the plastic reduction of the pulmonary artery volume with the Lecompte maneuver and the classical treatment of intracardiac changes in the heart [4]. The severity of symptoms in patients with tetralogy of Fallot and obstructive stenosis of the right ventricular outflow tract may vary. These patients are clinically divided into two groups: symptomatic and asymptomatic.

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The clinical manifestations determine the tactics of management of this group of patients. Patients may develop a variety of symptoms. The symptomatic group includes patients in whom this defect is manifested by episodes of desaturation, that is, a decrease in the concentration of oxygen in the blood. Most often, this symptom occurs in children due to anxiety or crying. The mechanism of action is due to the formation of severe obstruction in the right ventricular outflow tract. This obstruction can be dynamic or static. The mechanism of occurrence of these attacks is the development of dynamic muscular stenosis of the obstruction in the right ventricular tract, as a result of which the pressure in the right ventricle increases sharply and a significant part of the blood flow from the right ventricle flows into the left ventricle and into the aorta due to the intraventricular septal defect, while at the same time causing severe pulmonary hypoperfusion and cyanotic attacks due to a sharp decrease in the concentration of oxygen in the blood. The second subgroup in the symptomatic group of tetralogy of Fallot is formed by newborn patients in whom this heart defect manifests itself immediately after birth. In such patients, this occurs due to the closure of the patent ductus arteriosus, and the lungs are the main part of the circulatory system [6].

Epidemiology

Tetralogy of Fallot is diagnosed in 8-13% of all patients with congenital heart disease. Among the defects requiring surgical treatment in early childhood, tetralogy of Fallot accounts for 15%. The frequency of defects in newborns is from 4 to 7%. The average life expectancy of patients with tetralogy of Fallot is 7-8 years and depends on the degree of pulmonary artery valve stenosis. Mortality in the first year of life is 25%, at 3 years - 40%, at 10 years - 70%, at 40 years of life - 95% [9, 17, 18].

Tetralogy of Fallot is one of the most common cyanotic heart defects. Despite extensive experience in the treatment of this pathology, there is currently no generally accepted treatment strategy for patients with severe cyanosis, and the optimal age for radical surgical correction of tetralogy of Fallot in asymptomatic patients has not been determined [12, 15].

EchoCG

EchoCG allows you to determine the following anatomical components of this defect: the degree of pulmonary artery valve stenosis, the degree of aortic dextroposition, the size of the intraventricular septal defect, and the degree of right ventricular hypertrophy. Examination of the heart chambers reveals high pressure in the right ventricle, insufficient oxygen saturation of arterial blood, and the passage of a catheter from the right ventricle to the aorta.

ECG

Electrocardiography reveals rightward electrical shift of the heart, right ventricular myocardial hypertrophy, and incomplete right heart block [19].

Treatment of Tetralogy of Fallot through cardiac surgery

Currently, there is a global trend in pediatric cardiac surgery for early surgical treatment of congenital heart defects [14]. Newborns and young children with severe tetralogy of Fallot require palliative surgery at the initial stage, as well as to reduce the risk of complications in the subsequent treatment of this defect. The following types of palliative surgery are used to treat this defect:

- Use of Blalock-Taussing subclavian pulmonary anastomosis;
- Anastomosis between the ascending aorta and the right pulmonary artery;
- Central aortopulmonary anastomosis using a synthetic or biological prosthesis;

Anastomosis between the descending aorta and the left pulmonary artery.

Specific complications of surgeries used to treat tetralogy of Fallot may include: acute heart failure, pulmonary hypertension, right ventricular aneurysm, AV block, arrhythmias, and infective endocarditis.

Conclusion. Tetralogy of Fallot is a congenital heart defect, the pathological condition is characterized by the following four anomalies: intraventricular septal defect, right ventricular hypertrophy, aortic dextroposition, and pulmonary artery stenosis. This defect leads to dyspnea, cyanosis, and other serious complications due to insufficient oxygen supply to the heart. Treatment

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methods require cardiosurgical intervention. If the disease is diagnosed and treated as soon as possible, the patient's vitality can be maintained for a long time.

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