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CONGENITAL HEART DEFECTS IN CHILDREN

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Abstract:
Congenital heart defects are a group of diseases united by the presence of anatomical defects of the heart, its valvular apparatus or vessels that occurred in the prenatal period, leading to changes in intracardiac and systemic hemodynamics. The manifestations of congenital heart disease depend on its type; the most characteristic symptoms include pallor or cyanosis of the skin, heart murmurs, lag in physical development, signs of respiratory and heart failure. If a congenital heart defect is suspected, an ECG, **FCG**, radiography, EchoCG, cardiac catheterization and aortography, cardiography, MRI of the heart, etc. are performed. Most often, with congenital heart defects, cardiac surgery is resorted to – surgical correction of the detected anomaly.

Key words: congenital heart defects, epidemiology, prevalence, mortality, risk factors. General information

Аннотация: Врожденные пороки сердца – **группа заболеваний,** объединенных наличием анатомических дефектов сердца, его клапанного аппарата или сосудов, возникших во внутриутробном периоде, приводящих к изменению внутрисердечной и системной гемодинамики. Проявления врожденного порока сердца зависят от его вида; к наиболее характерным симптомам относятся бледность или синюшность кожных покровов, шумы в сердце, отставание в физическом развитии, признаки дыхательной и сердечной недостаточности. При подозрении на врожденный порок сердца выполняется ЭКГ, ФКГ, рентгенография, ЭхоКГ, катетеризация сердца и аортография, кардиография, МРТ сердца и т. д. Чаще всего при врожденных

пороках сердца прибегают к кардиохирургической операции – оперативной коррекции выявленной аномалии.

Congenital heart defects are a very extensive and diverse group of diseases of the heart and large vessels, accompanied by changes in blood flow, overload and heart failure. The incidence of congenital heart defects is high and, according to various authors, ranges from 0.8 to 1.2% among all newborns. Congenital heart defects account for 10-30% of all congenital anomalies. The group of congenital heart defects includes both relatively mild developmental disorders of the heart and blood vessels, as well as severe forms of heart pathology incompatible with life.

Many types of congenital heart defects occur not only in isolation, but also in various combinations with each other, which significantly aggravates the structure of the defect. In about a third of cases, cardiac abnormalities are combined with non-cardiac congenital malformations of the central nervous system, musculoskeletal system, gastrointestinal tract, genitourinary system, etc.

The most common types of congenital heart defects found in cardiology include ventricular septal defects (LVD – 20%), atrial septal defects (ATRI), aortic stenosis, aortic coarctation, open ductus arteriosus (OAP), transposition of large main vessels (SCV), pulmonary artery stenosis (10-15% of each

Causes of congenital heart defects

The etiology of congenital heart defects can be caused by chromosomal abnormalities (5%), gene mutation (2-3%), environmental factors (1-2%), polygenic-multifactorial predisposition (90%).

Various kinds of chromosomal aberrations lead to quantitative and structural changes in chromosomes. With chromosomal rearrangements, multiple polysystemic developmental abnormalities are noted, including congenital heart defects. In the case of autosomal trisomy, the most common heart defects are atrial or ventricular septal defects, as well as their combination; with sex chromosome abnormalities, congenital heart defects are less common and are mainly represented by aortic coarctation or ventricular septal defect.

Congenital heart defects caused by mutations of single genes are also in most cases combined with abnormalities of other internal organs. In these cases, heart defects are part of autosomal dominant (Marfan, Holt-Oram, Cruson, Noonan syndromes,

etc.), autosomal recessive syndromes (Kartagener, Carpenter, Roberts, Hurler syndrome, etc.) or X-linked syndromes (Goltz, Aase, Hunter syndromes, etc.).

Among the damaging environmental factors, viral diseases of pregnant women, ionizing radiation, certain medications, harmful habits of the mother, and occupational hazards lead to the development of congenital heart defects. The critical period of adverse effects on the fetus is the first 3 months of pregnancy, when fetal organogenesis occurs.

Intrauterine damage to the fetus by the rubella virus most often causes a triad of anomalies – glaucoma or cataracts, deafness, congenital heart defects (tetrad of Fallot, transposition of the main vessels, open ductus arteriosus, common arterial trunk, valvular defects, pulmonary artery stenosis, VHDL, etc.). microcephaly, impaired development of the bones of the skull and skeleton also usually occur. mental and physical retardation.

In addition to pregnant rubella, chickenpox, herpes simplex, adenovirus infections, serum hepatitis, cytomegaly, mycoplasmosis, toxoplasmosis, listeriosis, syphilis, tuberculosis, etc. pose a danger to the fetus in terms of the development of congenital heart defects.

The structure of embryophetal alcohol syndrome usually includes defects of the interventricular and atrial septum, and an open ductus arteriosus. It has been proven that the teratogenic effect on the fetal cardiovascular system is exerted by taking amphetamines, leading to transposition of the main vessels and LVEF; anticonvulsants that cause the development of aortic and pulmonary artery stenosis, aortic coarctation, open ductus arteriosus, Fallot's tetrad, hypoplasia of the left heart; lithium preparations that lead to tricuspid valve atresia, Ebstein's anomaly, DMPP; progestogens that cause Fallot's tetrad and other complex congenital heart defects.

In women suffering from prediabetes or diabetes, children with congenital heart defects are born more often than in healthy mothers. In this case, the fetus usually develops breast cancer or transposition of large vessels. A woman with rheumatism has a 25% chance of having a child with a congenital heart defect.

In addition to the immediate causes, there are risk factors for the formation of fetal heart abnormalities. These include the age of a pregnant woman under 15-17 years old and over 40 years old, toxicosis of the first trimester, the threat of spontaneous

termination of pregnancy, endocrine disorders in the mother, a history of stillbirth, the presence of other children and close relatives with congenital heart defects in the family.

Classification of congenital heart defects

There are several classifications of congenital heart defects based on the principle of hemodynamic changes. Taking into account the effect of the defect on pulmonary blood flow, the following are distinguished:

• congenital heart defects with unchanged (or slightly altered) blood flow in the small circulatory system: aortic valve atresia, aortic stenosis, pulmonary valve insufficiency, mitral defects (valve insufficiency and stenosis), adult-type aortic coarctation, atrial fibrillation, etc.

• congenital heart defects with increased blood flow in the lungs: not leading to the development of early cyanosis (open ductus arteriosus, DMPP, LVEF, aortopulmonary fistula, coarctation of the aorta of the pediatric type, Lutambache syndrome), leading to the development of cyanosis (tricuspid valve atresia with large LVEF, open ductus arteriosus with pulmonary hypertension)

• congenital heart defects with depleted blood flow in the lungs: not leading to the development of cyanosis (isolated pulmonary artery stenosis), leading to the development of cyanosis (complex heart defects – Fallot's disease, hypoplasia of the right ventricle, Ebstein anomaly)

• combined congenital heart defects, in which anatomical relationships between large vessels and various parts of the heart are disrupted: transposition of the main arteries, common arterial trunk, Taussig-Bing anomaly, divergence of the aorta and pulmonary trunk from one ventricle, etc.

In practical cardiology, congenital heart defects are divided into 3 groups: defects of the "blue" (cyanotic) type with a venoarterial shunt (Fallot triad, Fallot tetrad, transposition of the main vessels, tricuspid valve atresia); defects of the "pale" type with arteriovenous discharge (septal defects, open ductus arteriosus); defects with an obstacle to ways of blood ejection from the ventricles (stenosis of the aorta and pulmonary artery, coarctation of the aorta).

Hemodynamic disorders in congenital heart defects

As a result of the above-mentioned reasons, the correct formation of heart structures in the developing fetus may be disrupted, which is expressed in incomplete or untimely closure of the membranes between the ventricles and atria, improper valve formation, insufficient rotation of the primary heart tube and underdevelopment of the ventricles, abnormal vascular arrangement, etc. After birth, the ductus arteriosus and the oval window remain open in some children, which function physiologically in the prenatal period.

Due to the peculiarities of antenatal hemodynamics, the blood circulation of the developing fetus with congenital heart defects, as a rule, does not suffer. Congenital heart defects appear in children immediately after birth or after some time, which depends on the timing of the closure of communication between the large and small circulatory circles, the severity of pulmonary hypertension, pressure in the pulmonary artery system, the direction and volume of blood discharge, and the individual adaptive and compensatory capabilities of the child's body. Respiratory infection or some other disease often leads to the development of severe hemodynamic disorders in congenital heart defects.

With congenital heart defects of the pale type with arteriovenous discharge due to hypervolemia, hypertension of the small circulatory circle develops; with blue type defects with venoarterial shunt, hypoxemia occurs in patients.

About 50% of children with a large discharge of blood into the small circulatory system die without cardiac surgery in the first year of life from heart failure. In children who have crossed this critical threshold, blood discharge into the small circle decreases, well-being stabilizes, but sclerotic processes in the vessels of the lungs gradually progress, causing pulmonary hypertension.

In cyanotic congenital heart defects, venous discharge of blood or its mixing leads to overload of the large and hypovolemia of the small circulatory system, causing a decrease in blood oxygen saturation (hypoxemia) and the appearance of cyanosis of the skin and mucous membranes. To improve ventilation and perfusion of organs, a collateral circulatory network develops, therefore, despite pronounced hemodynamic disorders, the patient's condition may remain satisfactory for a long time. As the compensatory mechanisms are depleted, severe irreversible dystrophic

changes in the heart muscle develop due to prolonged myocardial hyperfunction. In cyanotic congenital heart defects, surgical intervention is indicated already in early childhood.

Symptoms of congenital heart defects

The clinical manifestations and course of congenital heart defects are determined by the type of anomaly, the nature of hemodynamic disorders, and the timing of circulatory decompensation.

Newborns with cyanotic congenital heart defects have cyanosis (cyanosis) of the skin and mucous membranes. Cyanosis increases with the slightest exertion: sucking, crying baby. White heart defects are manifested by paleness of the skin, coldness of the extremities.

Children with congenital heart defects are usually restless, refuse to breastfeed, and get tired quickly during the feeding process. They develop sweating, tachycardia, arrhythmias, shortness of breath, swelling and pulsation of the vessels of the neck. With chronic circulatory disorders, children lag behind in weight gain, growth, and physical development. With congenital heart defects, cardiac murmurs are usually heard immediately after birth. Later, signs of heart failure are detected (edema, cardiomegaly, cardiogenic hypotrophy, hepatomegaly, etc.).

Complications of congenital heart defects can include bacterial endocarditis, polycythemia, peripheral vascular thrombosis and cerebral vascular thromboembolism, congestive pneumonia, syncopal conditions, dyspnea, angina pectoris, or myocardial infarction.

Diagnosis of congenital heart defects

Detection of congenital heart defects is carried out through a comprehensive examination. When examining a child, the color of the skin is noted: the presence or absence of cyanosis, its nature (peripheral, generalized). Cardiac auscultation often reveals a change (weakening, amplification, or splitting) of heart tones, the presence of murmurs, etc. Physical examination in case of suspected congenital heart disease is complemented by instrumental diagnostics - electrocardiography (ECG), phonocardiography (FCG), chest X-ray, echocardiography (EchoCG).

An ECG can reveal hypertrophy of various parts of the heart, pathological deviation of the EOS, the presence of arrhythmias and conduction disturbances, which, together with data from other clinical examination methods, allows us to assess the severity of congenital heart disease. With the help of daily Holter ECG monitoring, hidden rhythm and conduction disturbances are detected. Through PCG, the nature, duration, and localization of heart tones and noises are assessed more thoroughly and in detail. Chest X-ray data complement previous methods by assessing the state of the small circulatory system, the location, shape and size of the heart, and changes in other organs (lungs, pleura, spine). During EchoCG, anatomical defects of the septa and valves of the heart, the location of the main vessels, and the contractility of the myocardium are visualized.

In complex congenital heart defects, as well as concomitant pulmonary hypertension, for the purpose of accurate anatomical and hemodynamic diagnosis, it becomes necessary to perform probing of the heart cavities and angiocardiography.

Treatment of congenital heart defects

The most difficult problem in pediatric cardiology is the surgical treatment of congenital heart defects in infants. Most operations in early childhood are performed for cyanotic congenital heart defects. If the newborn has no signs of heart failure or moderate cyanosis, the operation may be postponed. Children with congenital heart defects are monitored by a cardiologist and a cardiac surgeon.

The specific treatment in each case depends on the type and severity of the congenital heart defect. Operations for congenital defects of the septum of the heart (DVT, DMPP) may include plastic surgery or suturing of the septum, X-ray endovascular occlusion of the defect. In the presence of severe hypoxemia, children with congenital heart defects undergo palliative care at the first stage, involving the imposition of various kinds of intersystemic anastomoses. This tactic improves blood oxygenation, reduces the risk of complications, and allows for radical correction in more favorable conditions. In aortic malformations, resection or balloon dilation of aortic coarctation, plastic surgery of aortic stenosis, etc. is

performed. With OAP, it is bandaged. Treatment of pulmonary artery stenosis consists of open or endovascular valvuloplasty, etc.

Anatomically complex congenital heart defects, in which radical surgery is not possible, require hemodynamic correction, i.e. separation of arterial and venous blood flows without eliminating the anatomical defect. In these cases, the operations of Fontaine, Senning, Mustard, etc. can be performed. Serious defects that cannot be treated surgically require a heart transplant.

Conservative treatment of congenital heart defects may include symptomatic therapy of dyspnoea-cyanotic seizures, acute left ventricular failure (cardiac asthma, pulmonary edema), chronic heart failure, myocardial ischemia, and arrhythmias.

Prognosis and prevention of congenital heart defects

Congenital heart defects occupy the first place in the structure of newborn mortality. Without qualified cardiac surgery, 50-75% of children die during the first year of life. During the compensation period (2-3 years), mortality is reduced to 5%. Earlier detection and correction of congenital heart disease can significantly improve the prognosis.

Prevention of congenital heart defects requires careful pregnancy planning, exclusion of the effects of adverse factors on the fetus, medical and genetic counseling and awareness-raising among women at risk for having children with heart disease, addressing the issue of prenatal diagnosis of the defect (ultrasound, chorionic biopsy, amniocentesis) and indications for termination of pregnancy. Pregnancy management in women with congenital heart defects requires increased attention from an obstetrician-gynecologist and a cardiologist.

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