

The Structure of Cardiovascular Pathology in Childhood

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Article Information

Received: March 15, 2023

Accepted: April 13, 2023

Published: May 6, 2023

Keywords

aortic stenosis, aortic coarctation, pulmonary artery atresia, Ebstein anomaly.

ABSTRACT

The structure of cardiovascular pathology in childhood has undergone significant changes in recent decades. The decrease in the proportion of rheumatic diseases and infectious myocardial lesions occurs against the background of an increase in the number of cases of congenital heart defects, heart rhythm and conduction disorders, cardio-myopathies. Cardiovascular pathology is one of the main causes of mortality regardless of age. Neonatal mortality prevails in the structure of infant mortality, accounting for more than 60% of all deaths in the first year of life.

Congenital malformations occupy the second place among the causes of infant mortality. Congenital heart and cardiovascular system defects account for 47% (12.2 per 10 thousand live births) of all causes of death from malformations. The total mortality in congenital heart defects is extremely high, by the end of the 1st week 29% of newborns die, by the 1st month - 42%, by the 1st year - 87% of children [1]. It is obvious that the possibility of providing assistance with this pathology is determined by its detectability in the first days of life.

The modern level of diagnosis and treatment in most cases allows you to save the lives of children with congenital heart defects. Critical defects account for approximately 25% of all congenital heart defects. Thus, every year about 5 thousand children are born with heart defects, in which surgical correction is indicated in the first days of life. However, according to the published information, surgical care in the neonatal period is provided to 2-2.5 thousand newborns [2]. Consequently, further improvement of medical care for patients with congenital heart defects should be largely aimed at improving pre/postnatal diagnosis, optimizing the timing and methods of surgical correction, developing issues of nursing and rehabilitation of newborns after cardiac surgery.

Significant advances in neonatology, the expansion of diagnostic capabilities in general, the development of pediatric cardiology and cardiac surgery have led to a significant increase in the number of children with complex congenital heart defects who live to the age of 18. At the same time, the vector of further development of cardiology is increasingly shifting towards the fetus and the newborn child. The definition of the concept of "perinatal cardiology" seems extremely relevant, primarily due to the indicated significance of congenital heart pathology.

Understanding the causes of the formation of congenital pathology of the cardiovascular system, the basics of hemodynamics in the fetus and newborn child allows not only to diagnose the existing problem in a timely manner, but also to provide therapeutic and surgical assistance.

Prenatal diagnosis of congenital heart defects ensures timely transfer of a child to a cardiac surgery hospital for a critical defect or to a specialized cardiology department for dynamic observation and treatment. The widespread introduction of fetal echocardiography into the practice of monitoring pregnancy has significantly increased the detection of complex and critical forms of heart defects. Despite the intensive development of this service, an average of 34-38% of heart defects are diagnosed antenatally [3, 4]. The qualification and experience of a doctor are of enormous importance in the adequate diagnosis of complex heart defects. The anomaly of the number of chambers of the heart is detected much more often and better than pathology at the level of the ventricular excretory divisions and main vessels.

However, the accuracy of prenatal diagnostics is determined not only by the qualification of a specialist and the availability of expert-class equipment, but also by what positions are used to examine the structures of the heart with two-dimensional echocardiography (EchoCG). So, in the study of H. Liu et al. [5] the sensitivity of the prenatal diagnostic method was 68.1%, and the specificity was 99.9%. It is shown that the sensitivity of the method when examining only a four-chamber projection of the fetal heart is 48.7%, when examining a four-chamber projection of the fetal heart and ventricular outflow - 58%, and when examining a four-chamber projection of the fetal heart, ventricular outflow and projection of 3 vessels - 77.1%. In the work of M.C. Rossier et al. [6] complex congenital malformations (variants of the single ventricle of the heart) were diagnosed in utero in 81% of cases, transposition of the main arteries – in 67%. At the same time, heart defects requiring follow-up and treatment after birth (aortic stenosis, aortic coarctation, pulmonary artery atresia and Ebstein anomaly) were detected only in 26% of cases [6]. This group of defects is accompanied by the rapid development of hemodynamic disorders, and the lack of timely pre- and postnatal diagnosis determines a high probability of death in the early neonatal period. Currently, in our country, the current protocol of ultrasound examination of the fetus at the 18th-21st week of pregnancy includes only verification of a four-chamber section of the heart. The change of the specified protocol towards the mandatory examination of the excretory sections of the ventricles of the heart and the cut through 3 vessels will significantly increase the detection of congenital heart defects.

There are a significant number of classifications of congenital heart defects, the most important of which is the separation of defects in terms of the criticality of the situation in the newborn period. It should be noted that the concept of critical heart defect after birth is often replaced in practice by the term "ductus-dependent defect". However, a critical congenital heart defect of the newborn period is considered to be a defect that depends on the functioning of any fetal communications (both an open arterial duct and an open oval window). Heart defects that depend on the functioning of an open arterial duct are called ductus-dependent, and foramen-dependent on the functioning of an open oval window.

The regulatory document defining the routing of a pregnant woman with a fetal heart defect in our country, depending on the timing of operative correction of the defect, is the order of the Ministry of Health of the Russian Federation dated 01.11.2012 No. 572n (ed. dated 17.01.2014). The order clearly states the timing of an ultrasound examination of a woman during pregnancy. Ultrasound screening at the 18th-21st week of pregnancy is designed to identify "late-manifesting congenital anomalies of the fetus." A number of heart defects can become

critical during intrauterine development, and in the case of prenatal detection of congenital heart disease at 20 weeks, a second examination of the fetus is required at least at 35-36 weeks.

The detection of foramen-dependent heart defects determines the need for expert fetal echocardiography at least 2 times during pregnancy and preferably at 37-38 weeks to predict the need for emergency atrioseptomy surgery immediately after the birth of the child. The group of congenital heart defects, in which an emergency transfer to a cardiosurgical hospital is indicated, includes most of these defects, primarily ductus-dependent. However, the order does not contain the concept of "form- dependent defects", in which the territorial proximity of a cardiac surgical hospital or the possibility of performing an emergency cardio- surgical operation in a perinatal center is vital.

In addition, one of the most fatal congenital heart defects – an abnormal departure of the left coronary artery from the pulmonary trunk – is attributed to the group of congenital heart defects in which a planned surgical intervention is required . The same group includes a hemodynamically significant open arterial duct in the unborn, which is not a heart defect in this category of patients. A hemodynamically significant open arterial duct may pose a threat to the life of a child with an extremely low birth weight and in some cases necessitates emergency surgical correction. In the group of congenital heart defects requiring intervention in the first 3 months of life, heart defects are attributed, the evolution of which in the process of intrauterine development can lead to a critical condition of the child right after birth. In this case, the delivery of a pregnant woman should be carried out in an obstetric hospital that is not geographically connected with a cardiosurgical center. Most newborns do not need specialized cardiac surgery in the first hours after birth. The probability of developing a particular condition is based on postnatal observations. Prenatal diagnosis of a heart defect, in which emergency specialized care is required after birth or at the time of cord clamping, determines the necessity of planning the birth of such a patient.

According to the recommendations of the American Association of Cardiologists (AHA) on the diagnosis and treatment of fetal heart disease, the type of heart defect, its severity, the presence of intrauterine decompensation of blood circulation and arrhythmia, as well as concomitant developmental abnormalities determines the management tactics of a pregnant woman [7]. In addition, these recommendations take into account foramen-dependent heart defects, define a list of therapeutic and diagnostic procedures that a child may need immediately after birth, a list of specialists to perform them and the method /term of delivery of a woman in the interests of the fetus.

The key to successful postnatal treatment of a child with congenital heart disease is not only in timely intrauterine diagnosis of the defect, but also in the proper management of childbirth and the early neonatal period. Only the coordination of the work of gynecologists, neonatologists, cardiologists with the provision of intensive therapy, emergency interventional or surgical intervention, if necessary, allows us to fully count on a good prognosis. The available data indicate that the delivery solution in the immediate vicinity of the cardio- surgical hospital significantly improves the condition of the child during the newborn period and the results of surgical intervention in cases of critical congenital heart defects.

The probability of a catastrophic violation of circulation after birth with a detected heart defect makes it necessary to plan further pregnancy management and a clear scheme of actions of medical personnel at the time of delivery. Practice has shown that prolonging pregnancy to 39 weeks improves the survival rate of new- born children with congenital heart disease. It is not

recommended to make a decision on the pre- temporary stimulation of labor activity, if it is not about vital indications for the condition of the mother or fetus.

To date , the necessity or potential advantages of cesarean section surgery in women in the detection of fetal heart disease have not been convincingly proven (with the exception of congenital heart defects with intrauterine decompensation of blood circulation to increase the chance of survival) [8]. Nevertheless, the data of published studies suggest that the decision to deliver through the natural birth canal in most cases is made in the absence of information about the presence of congenital heart disease in the fetus.

Recommendations on the method of delivery of a woman with a fetal heart defect should find their place in the following editions of the directive documents on the provision of medical care to patients of this category. Timely prenatal diagnosis of congenital heart pathology allows not only to provide adequate therapeutic and surgical care, but also to minimize complications arising at any stage of treatment of a child after birth. Postnatal diagnosis of heart defects is extremely important from the point of view of the large contribution of this problem to the structure of early neonatal and infant mortality. In this aspect, it is extremely important to identify critical forms of heart defects of the newborn period, which determine mortality in the first day and weeks after birth.

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