

## CONGENITAL HEART DISEASE

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**Abstract.** Congenital heart disease (CHD) is a defect in the structure of the heart and large vessels diagnosed from the moment of birth. The frequency of occurrence varies widely and ranges from 2.4 to 14.15%. Every year, more than 30,000 children with congenital heart defects are born worldwide. For one million of the adult population, there are about 3,500 people with this pathology. Many of these patients are young women planning to carry a pregnancy and give birth to a healthy child.

**Keywords:** Turner syndrome, Down syndrome, Williams syndrome, Mendel syndrome.

In Western countries, congenital heart defects are the most common form of cardiovascular diseases that complicate the course of pregnancy and childbirth. 74% of cases of complications were registered according to the Canadian CARPREG registry and 66% of complications according to The European Register ROPAC [3,4]. In women with congenital heart defects during pregnancy, the risk of developing such severe complications as heart failure, arrhythmias, cerebrovascular diseases, embolisms up to a fatal outcome increases. Prolonged overload of the ventricle of the heart by volume and pressure is possible due to damage to the heart valve or the presence of an incorrect shunt, especially with late surgical correction or palliative surgical defects. The development of arrhythmias, most often supraventricular, is associated with dilation of the chambers of the heart, leading to myocardial hypertrophy, fibrosis, scarring and disorders of the conduction system. Pregnancy can provoke the development of arrhythmias on a healthy heart due to such physiological features as hormonal exposure, physiological dilation, and changes in the autonomic nervous system [7]. Mothers with congenital heart defects are often diagnosed with fetal growth retardation, newborns have a lower birth weight, are more often born prematurely, and have a risk of developing metabolic and cardiovascular complications. Parents with congenital heart disease have a high risk of developing defects in children. The risk of developing heart disease in an unborn child increases to 10% in case of the presence of this pathology in a mother or in one relative in a generation [3,8]. In patients with congenital heart defects, specific genetic anomalies are diagnosed, such as Turner, Down, Williams, Mendel syndrome with mutation in one gene, cyclocardiofascial syndrome, syndrome Noonan, Holt-Oram syndrome, heterotaxial syndrome, which significantly complicates their management. With Marfan syndrome or other autosomal dominant syndromes, the risk of transmission of a heart anomaly to the fetus is 50%. Open ductus arteriosus, tetrad Fallo, transpositional defects can be associated with chromosomal abnormalities, such as trisomy 21, 13 or 18 chromosomes or with microdeletion 22q11.2. 10-15% of cases of Fallo tetrad development and pulmonary artery atresia are associated with the latter anomaly, while the risk of transmission of the defect to offspring is very high. Such somatic diseases as diabetes mellitus and obesity, smoking, increase the risk of developing congenital heart disease of the fetus several times. In turn, folic acid intake at the stage of preliminary preparation and during pregnancy reduces the risk of the formation of congenital heart disease in the unborn child [9, 10]. Young women have insufficient information about their existing heart disease, the

possibility of pregnancy, the complications of the gestational period, childbirth and the main disease. In this regard, information support aimed at highlighting the issues of pregnancy prevention in this group. Based on the risk assessment of pregnancy and childbirth in women with congenital heart defects, the American Heart Association (ACC/ANA), the American Society of Cardiologists and the European Society of Cardiology (ESC) have developed recommendations for the optimization of the management of this group of patients [7]. When assessing risk factors in pregnant patients with congenital heart defects, it is necessary to take into account the anatomical features of the defect, the type of surgical intervention performed earlier, to assess the functional capacity of the heart based on the data of anamnesis and stress tests. One of the main factors of the unfavorable outcome of pregnancy and childbirth in women with congenital heart defects is the presence of cardiac insufficiency, which is diagnosed when performing a test with physical activity [12]. To diagnose the topic, functional ability and features of hemodynamics of the heart, it is necessary to conduct additional research methods: electrocardiography, echocardiography with an assessment of the functions of the ventricles, heart valves, the degree of pulmonary hypertension, the material from which the prostheses and patches are made. In pregnant women with congenital heart defects, computer and magnetic resonance imaging of the heart, catheterization of the heart cavities are performed, the concentration of cardiac bio-markers is determined. All patients with congenital heart defects who are still planning pregnancy, it is mandatory to recommend genetic examination and consultation of a geneticist [13]. It was revealed that in the population the most widespread congenital heart defects are ventricular rupture defect - the frequency of occurrence is 27-42%; atrial septal defect - 5-15% of cases; open ductus arteriosus - in 10-18%; aortic coarctation - 7% of cases; congenital aortic stenosis - 6%; pulmonary artery stenosis - 8-10%; Fallot group defects - 5% [1]. Hemodynamic disorders and clinical manifestations in congenital heart defects vary depending on the size of the defect, localization, nature and duration of the defect [7]. Pregnancy itself places very high demands on the cardiovascular system of the body, especially in women with congenital heart defects, whose compensatory capabilities are already reduced. The main clinical symptoms in pregnant women with heart defects do not have a specific character. Worries about rapid fatigue, muscle weakness, heaviness in the legs, a feeling of drowsiness, palpitations and shortness of breath that occur during physical exertion. With the progression of hemodynamic disorders in congenital heart disease, shortness of breath is observed at rest [4]. Congenital heart defects are one of the main causes of maternal mortality during pregnancy and require accurate diagnosis, detailed monitoring during pregnancy together with cardiologists and cardiac surgeons, properly selected medical and surgical correction. In the structure of maternal mortality, heart diseases account for 28.5%. In pregnant women with congenital heart defects, blood oxygen saturation decreases, circulatory or mixed respiratory-circulatory hypoxia develops. The existing chronic hypoxia of the mother leads to a violation of the uteroplacental blood flow and, as a consequence, to chronic hypoxia of the fetus [7,9]. Thanks to cardiac surgery and interventional cardiology, many women with congenital heart defects reach childbearing age, plan and carry pregnancy [13]. The Working research Group of the European Society of Cardiology proposed to use the unified modified WHO classification for assessing the risk of cardiovascular complications for mother and unborn child in pregnant women with congenital heart defects and other cardiovascular pathology. The proposed classification combines all cardiovascular risk factors, underlying heart disease, concomitant diseases, includes possible contraindications to pregnancy, the timing of consultations of narrow specialists,

additional research methods, the timing and methods of resolution depending on the type of congenital malformation and the presence of complications [7]. The modified WHO classification of maternal risk in cardiovascular diseases includes several groups of patients [7]. The first group includes conditions in patients with WHO risk assessment I: uncomplicated, minor or moderately pronounced pulmonary artery stenosis and open arterial duct; successfully operated heart defect with atrial peritoneal defect, ventricular peritoneal defect, open ductus arteriosus, abnormal pulmonary drainage veins, rare pre-cardiac or ventricular extrasystoles. The second group includes conditions in patients with WHO risk assessment II or III. WHO II includes: an unoperated defect of the interventricular septum and a defect of the interventricular septum, a corrected tetrad Fallo, almost all arrhythmias in the absence of other complications. WHO III includes: moderate lesion of the left ventricle, hypertrophic cardiomyopathy, diseases of the valvular apparatus (not corresponding to WHO I or IV), Marfan syndrome, not complicated by aortic dilation, aortic diameter less than 45 mm in combination with a bicuspid aortic valve, operated aortic coarctation. WHO III without taking into account individual characteristics – artificial mechanical valve, systemic right ventricle, Fontaine surgery, non-operated heart defects with cyanosis, other complex congenital heart defects, dilation of the aorta 40-45 mm with Marfan syndrome, dilation of the aorta 45-50 mm with a bicuspid aortic valve. The third group includes conditions in pregnant women with WHO risk assessment IV, in which pregnancy is contraindicated. These include pulmonary arterial hypertension, systemic ventricular dysfunction with a withdrawal fraction of less than 30% and functional class III-IV heart failure, pre-existing periparturient cardiomyopathy with residual lesion of the left ventricle, pronounced mitral stenosis, pronounced aortic stenosis with subjective symptoms, Marfan syndrome with dilation aorta greater than 45 mm, dilation of the aorta more than 50 mm with a bicuspid aortic valve, pronounced aortic coarctation. Diagnosis of congenital heart defects includes not only anamnestic indications for the presence of a defect and heart murmurs from birth or childhood, but also a physical examination. Palpation of the heart area, percussion of the heart and vascular bundle, auscultation of the heart tones are required. Laboratory tests are carried out at the stage of pre-pregnancy preparation at the terms from 10 to 11, from 26 to 28 and at 32 weeks of gestation, as well as after delivery, and must necessarily include an assessment of the state of the blood coagulation system. Electrocardiography of the heart makes it possible to identify signs of hypertrophy and overload of various parts of the heart, depending on the type of defect and the existing hemodynamic disorders. Doppler echocardiography and echocardiography of the heart in most cases make it possible to detect signs characteristic of heart defects, assess the degree and severity of violations of intracardiac hemodynamics and the functional state of all parts of the heart [7,9,12]. Most often, congenital heart defects accompanied by pulmonary hypertension lead to the development of such severe heart failure. The frequency of maternal mortality in this group is the highest – 30-50%. This group also includes patients with severe aortic and aortic valve stenosis. Pregnancy is not recommended for such patients. In the case of its onset, an interruption is indicated due to the high risk of fatal complications in the mother. The issue of termination of pregnancy is decided by a consultation with the mandatory presence of cardiologists and cardiac surgeons. Even the termination of pregnancy itself is dangerous, as it is associated with a high risk of complications due to increased vasodilation and a decrease in myocardial contractility as a result of the anesthesia methods used [7,12]. The low-risk group includes pregnant women without pulmonary hypertension, with moderate valve insufficiency. With such defects, decompensation of cardiac activity during

pregnancy does not occur due to a decrease in total peripheral vascular resistance. With minor stenosis or moderate aortic stenosis, women also tolerate pregnancy well, in parallel with the increase in shock volume, the pressure gradient increases. A moderate degree of pulmonary artery stenosis is most often also well tolerated, occasionally surgical intervention is required during pregnancy. Patients with corrected heart defects without artificial valves do not tolerate the course of pregnancy. Residual defects after cardiac surgery occur in 2-50% of cases and should be confirmed clinically and during echocardiography. Special cardiological examination of patients of all groups is carried out every trimester. All pregnant women with congenital heart diseases need the constant supervision of a cardiologist and an obstetrician-gynecologist. Once a month, it is necessary to perform an echo cardioscopy, three times during pregnancy, hospitalization in a multi-profile hospital with a cardiologist in its composition is indicated. The first hospitalization is carried out in time up to 12 weeks of gestation in order to clarify the diagnosis, the functional state of the cardiovascular system, the presence and activity of the rheumatic process, solving the question of the possibility of pregnancy. The second hospitalization should be carried out in the period from 27 to 32 weeks of pregnancy, during the period when the cardiovascular system is experiencing the greatest load. In addition to a thorough examination, the patient is undergoing correction of the therapy. The last hospitalization is carried out two to three weeks before the due date in order to prepare for childbirth, resolve the issue of the timing and method of delivery, prescribe or correct cardiac and anti-rheumatic therapy. All additional hospitalizations of pregnant women with congenital heart defects should be justified by those who have joined obstetric and perinatal complications, as well as decompensation of the underlying disease [7]. In the presence of a congenital heart defect corresponding to the first degree - a very low risk of complications, pregnant women should be consulted by a cardiologist no more than twice during pregnancy. In the second degree (low or moderately high risk), a cardiologist is monitored every trimester of pregnancy. At the third degree (high risk of complications), a joint examination by a cardiologist and an obstetrician-gynecologist is carried out once every one to two months. The same management tactics are followed at the fourth degree of risk of complications if a woman refuses the decision recommended to her by the council on termination of pregnancy. If necessary, examinations are carried out more often [12,14]. Pre-pregnancy preparation plays an important role in the successful outcome of pregnancy for the mother and the unborn child in women with congenital heart defects. It includes valvuloplasty, therapeutic or surgical treatment of arrhythmias, treatment of concomitant diseases. The issue of prescribing anticoagulant therapy in pregnant women with a mechanical prosthetic heart valve must be resolved. Doctors should recommend the patient the most favorable period for pregnancy, depending on the type of congenital defect, its compensation, and the absence of severe complications. It is advisable to carry a pregnancy at the age of 20 to 25 years [2,4,5]. Despite the common approaches to the management of pregnancy in congenital heart defects, there are peculiarities depending on the type of defect and the presence of complications. With a defect of the interventricular septum, the management of pregnancy and childbirth depends on the high or low type of defect. The low type of ventricular septal defect has practically no hemodynamic disorders, proceeds favorably. Gestation and childbirth with this type of defect are not contraindicated. During pregnancy, a double visit to a cardiologist is enough. Delivery is carried out through the natural birth canal. With a severe defect of the interventricular septum, management tactics depend on the stage of heart failure and the degree of pulmonary hypertension. With minor and moderate pulmonary hypertension and heart failure of

the first stage, pregnancy is not contraindicated. Independent work is possible with the exception of a busy period. With severe pulmonary hypertension and the presence of symptoms of heart failure of the third and fourth functional class, pregnancy is contraindicated. In case of refusal of the patient from termination of pregnancy, it is indicated to be in a hospital during the entire gestational period. In the absence of pulmonary hypertension, delivery is performed by caesarean section. In the postpartum period, it is possible to develop such a complication as paradoxical systemic embolism [2,4,6,7]. Atrial septal defect is the most common congenital heart defect. According to clinical data, it occurs with a frequency of 5-15%. According to the pathoanatomical conclusions - 3.7–10% [1]. There are primary and secondary defects of the interstitial septum. Secondary occurs more often – in 95%. Pregnancy, as a rule, takes place well.

Moderate pulmonary hypertension in these patients is hypervolemic in nature and does not adversely affect the course of pregnancy. A hemodynamically significant defect of the atrial peritoneum should be closed before pregnancy. The course of the defect is complicated by thrombembolism and arrhythmias. Delivery is carried out through the natural birth canal. With an uncomplicated secondary defect of the atrial septum, pregnancy and childbirth are not contraindicated. In the presence of complications, tactics depend on their nature and severity. A contraindication to pregnancy is the presence of high pulmonary hypertension or Eisenmenger syndrome. In case of pregnancy, inpatient monitoring and medical correction of heart failure is recommended. Delivery is possible through the natural birth canal with the exception of the forced period. Delivery by caesarean section is performed with high pulmonary hypertension. The course of pregnancy is most often complicated by the development of preeclampsia and delayed fetal development. During the observation of pregnancy, a double examination by a cardiologist is sufficient. Secondary defects of the atrial junction are subject to catheter closure by Amplatzer in case of deterioration. In order to prevent paradoxical pulmonary embolism, it is not recommended to close minor defects of the atrial peritoneum, an open oval window. To prevent pulmonary embolism in chronic venous insufficiency of the legs, it is recommended to wear elastic tricotage, prolonged bed rest and the use of heparin [2,4,6,7]. Atrioventricular communication is characterized by the presence of communication between the atria and ventricles caused by defects in the septa, in combination with the opening of the mitral and tricuspid valves. There is an incomplete form of atrioventricular communication, which includes a defect of the atrial fibrillation with splitting of the mitral valve leaf, and a complete form, which is an almost single channel, including a defect of the atrial septum, a defect of the interventricular septum, the absence or splitting of the septal valves of the mitral and tricuspid valves. Such a defect is a single atrioventricular orifice. The full form of atrioventricular communication is often combined with Down syndrome and other heart defects. Signs of heart failure are characteristic for patients with such a defect from birth. It is practically impossible to reach the fertile age [2,4,6,7]. Open ductus arteriosus accounts for 10-18% of all congenital heart defects [1]. With an uncomplicated open arterial duct, pregnancy is not contraindicated. A contraindication to pregnancy is the addition of pulmonary hypertension [2,4,6,7]. Aortic coarctation occurs in 7% of all congenital heart defects, and may occur in Turner syndrome [1,10]. The question of the possibility of pregnancy is resolved only after surgical correction of the defect. Pregnancy is possible with moderate narrowing of the aorta and a blood pressure level of no more than 160/90 mm Hg. Due to the high risk of rupture of the altered aortic wall, delivery is performed by caesarean section. With mild aortic coarctation, it is possible to carry a pregnancy without surgical correction, but the likelihood of complications

due to arterial hypertension is high. Maternal mortality in this type of defect reaches 3.5%. During pregnancy, medical correction of arterial hypertension is carried out, cardioselective drugs are prescribed beta-blockers. With high figures of arterial pressure, cardiac insufficiency of functional class III-IV, violation of cerebral circulation, pregnancy is contraindicated [2,4,6,7]. During pregnancy, it is recommended to limit physical exertion, often hospitalization is carried out for the entire period of gestation. Due to the high risk of aortic dissection and rupture, balloon angioplasty and stenting are not performed [2,7]. Tactics in operated patients depends on the period of limitation of the operation, its type and effectiveness. With the duration of the operation less than a year, with the storage of high blood pressure, delivery by caesarean section is indicated, in other cases, the termination of the labor period [7]. Possible complications: cerebral hemorrhage, dissection and rupture of the aorta, infectious endocarditis, severe pre-eclampsia, spontaneous miscarriage, premature birth, delayed fetal development. In childbirth, epidural anesthesia is preferable. Pregnancy monitoring includes regular monitoring of blood pressure, daily monitoring of blood pressure every trimester, adequate hypotensive therapy. With high arterial hypertension, which is not amenable to medical correction, percutaneous plastic surgery of aortic coarctation can be performed [6,7]. Congenital aortic stenosis occurs in 6% of cases [1]. The vice is well tolerated for a long time. Due to the development of relative insufficiency of the mitral valve, hypertrophy of the left ventricle, its dilation and "mitralisation" develop over time. By itself, pregnancy can cause decompensation of the compensated defect. There are signs of relative coronary insufficiency, manifested by angina attacks and the development of myocardial infarction. Pregnancy and childbirth are possible with mild to moderate aortic stenosis, however, the risk of complications is very high. Thus, pregnancy with this defect is not viable, the decision on planning and gestation is decided individually by a consultation. The appearance of brain symptoms in the early stages, pain behind the sternum is an indication for termination of pregnancy. In severe aortic stenosis, a woman needs to explain the deadly danger associated with pregnancy and childbirth. Complications of heart disease: acute coronary insufficiency, acute left ventricular insufficiency, chronic heart failure, fatal rhythm and conduction disorders, acute cerebrovascular accident, sudden death [2,4,6,7]. Stenosis of the mouth of the pulmonary artery occurs in 8-10% [1]. With mild to moderate severity of stenosis, pregnancy and childbirth proceed without complications. In severe stenosis, pregnancy can contribute to the development of supraventricular arrhythmias and right ventricular failure. Surgical correction should be performed before pregnancy. In case of right ventricular failure with severe stenosis during pregnancy, balloon valvuloplasty is performed [2,4,7].

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