

### Ebstein's Anomaly

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#### ABSTRACT

Ebstein's anomaly is a fairly rare congenital heart defect. The frequency is 1%. Patients with Ebstein's anomaly without heart failure and cyanosis are undergoing pregnancy well. In patients with symptoms of heart failure, pronounced tricuspid regurgitation and cyanosis, surgical correction should be performed before pregnancy. If surgical intervention is refused, pregnancy is contraindicated.

Complications during pregnancy are directly related to the degree of tricuspid insufficiency and the functional ability of the right ventricle, combination with WPW syndrome and atrial septal defect, the risk of paradoxical embolization increases. During pregnancy, the frequency of premature birth, delayed fetal development, and perinatal mortality increases. Patients with tricuspid insufficiency or cardiac insufficiency should be monitored by a cardiologist at least once a trimester.

Childbirth through the natural birth canal is possible [4,7]. Transposition of the main vessels occurs with a frequency of 7-15% of all congenital heart defects [1]. Gestation of pregnancy is possible only with corrected transposition. A successful outcome of pregnancy is possible in 60% of cases. With a complete transposition of the main vessels, a Senning or Mustard operation is performed. With hemodynamic correction, the prognosis of pregnancy is favorable [7]. If, in the case of correction of the defect, the function of the right ventricle is impaired, heart failure of the third and fourth functional class develops, tricuspid insufficiency, the ejection fraction decreases by less than 40%, pregnancy is contraindicated and associated with a high risk of arterial hypertension and preeclampsia.

Women with a corrected transposition of the main vessels need to be examined by a cardiologist on a monthly basis, including echocardiography and daily monitoring of the Holter electrocardiogram. In the absence of symptoms of heart failure, satisfactory function of the right ventricle, childbirth through the natural birth canal is possible. With a decrease in the contractility of the heart, it is advisable to deliver by cesarean section [7]. With a single ventricle of the heart, there is no interventricular septum, and the structure of the heart is three-chamber.

When such a defect is combined with pulmonary artery stenosis, Fontaine surgery is indicated [1,2,3,7].

Fontaine's operation allows you to endure pregnancy up to a period of 25-35 weeks. Even in the case of surgery, the risk of maternal mortality is 2%, arrhythmias develop in 20%, enteropathy, hepatomegaly, cirrhosis of the liver are possible, the tendency to thrombosis and heart failure increases, the risk of premature birth and the birth of children with low body weight increases [12]. Pregnancy is contraindicated with a decrease in the contractile function of the right ventricle, oxygen saturation of less than 85% in some cases, moderate and pronounced atrioventricular regurgitation and protein enteropathy [7].

When carrying a pregnancy, monthly monitoring by a cardiologist is necessary. Due to the high risk of shunt thrombosis and thromboembolic complications, anticoagulants are prescribed, early delivery is performed by caesarean section [7]. The greatest practical significance among the vices of the Fallot group is the Fallot tetrad. The presence of a "blue" defect is a contraindication for carrying a pregnancy. With uncorrected malformation, maternal mortality is 7%, perinatal mortality reaches 22% [4]. Pregnancy proceeds well only after surgical correction of the defect. In 12% of cases, complications such as arrhythmias, heart failure, thrombolysis, progressive dilation of the aortic root, endocarditis occur during pregnancy.

In the presence of clinical signs of pronounced dilatation of the right ventricle, prosthetics of the valve should be performed before the planned pregnancy [2,4,6,7]. During pregnancy, a cardiologist's examination in each trimester is sufficient. Echocardiography is performed once a month. With the development of right ventricular insufficiency, diuretics and bed rest are indicated. In the absence of the effect of conservative surgery, catheter implantation of the valve or premature surgical delivery is performed [7]. In all cases, the preferred method of delivery is considered to be delivery through the natural birth canal. The danger of childbirth and the postpartum period is due to the fact that when the uterus contracts, venous blood rushes to the heart, but due to stenosis of the mouth of the pulmonary artery, it cannot pass completely into the small circle and its significant part through the ventricular septum defect enters the large circulation circle, dramatically increasing the already existing hypoxemia.

Thorough monitoring of blood pressure and blood gases is necessary during childbirth and the postpartum period. It is extremely important to prevent further vasodilation, including those induced by medicinal products. Syncopal, thromboembolic conditions, infectious endocarditis and sudden death are possible in such patients [4,15]. All patients with the Fallot tetrad undergo genetic examination to establish the deletion syndrome of chromosome 22q 11 using fluorescent hybridization in situ [7,9]. Eisenmenger's syndrome is called severe irreversible pulmonary hypertension with blood discharge in two directions or spread to the left through an open arterial passage, a defect of the atrial and interventricular septum. The formed Eisenmenger syndrome cannot be surgically corrected. The frequency of maternal mortality is very high – 30-50%, perinatal mortality is 28%.

During pregnancy and in the postpartum period, due to fibrinoid necrosis or pulmonary thrombosis, pulmonary vascular resistance increases, which leads to a fatal outcome. Due to systemic vasodilation and overload of the right ventricle with increased cyanosis and decreased blood flow through the pulmonary vessels during pregnancy, blood shunting from right to left increases. Epidural anesthesia is contraindicated in this group of patients. Complex and fatal arrhythmias may develop, and the risk of thromboembolic complications is high. With this defect, termination of pregnancy in the first trimester is recommended. If the patient refuses to interrupt pregnancy, hospitalization for the entire gestation period in a specialized institution is indicated [5,7].

The prognosis depends on the severity of pulmonary hypertension. The following therapeutic

and preventive measures are carried out: oxygen and bed rest for shortness of breath attacks, the use of anticoagulants from the second trimester and up to two days after childbirth [12,15]. In the second and third trimesters, anticoagulants are prescribed 3-4 weeks before the delivery date - heparin or low-molecular-weight heparins. With the exception of hemoconcentration and a decrease in the volume of circulating blood, small doses of diuretics can be prescribed with caution [15]. Independent childbirth with the exclusion of attempts is possible. During childbirth, monitoring of all body functions is carried out, three-fold determination of the level of blood gases, indicators of central and peripheral hemodynamics. Early delivery under regional anesthesia is indicated when the condition of the mother and fetus deteriorates. With acid saturation of less than 85%, the number of children born alive is insignificant [7,9].

Thus, there is currently an increase in patients with congenital heart defects. The number of women with pathology of the cardiovascular system planning and carrying a pregnancy does not decrease. Pregnant women with low-risk congenital heart defects can be observed by cardiologists of the general polyclinic network, while patients with more complex heart defects should be observed by cardiologists specializing in congenital heart defects. The American Council of Medical Specialists proposed to monitor pregnant women with congenital heart defects to a specially formed group of cardiologists with specialization in this field of cardiology and cardio-surgery. Doctors observing such patients should be high-class experts in the diagnosis and management of women with various specific congenital heart defects. Patients with congenital heart defects very often stop regular observation by doctors with a satisfactory state of health,

However, it is necessary to remember that when planning pregnancy in this group of patients, they need medical supervision. They may fall into a group of high-risk patients who need the supervision of highly qualified specialists. To optimize the care of pregnant women with congenital heart defects and achieve positive results, it is necessary to interact with doctors of different specialties: obstetricians, gynecologists, therapists, cardiologists, cardiac surgeons and specialists in congenital heart defects.

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