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**Features of Prenatal Diagnosis and Perinatal Management in Patients with Taussig-Bing anomaly**

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In the article it was analyzed the perinatal results of diagnosis and treatment of 24 fetuses with Taussig-Bing anomaly from 2006 to 2015. The majority of these patients (90.5%) were born in Kyiv near the cardiac center for consultations of the newborns on the first day of life to determine surgical treatment. The dominant concomitant cardiac anomaly in this group of patients was pathology of the aortic arch (to 57.1%), which only in half of the cases were diagnosed prenatally. Postnatal finding was a high percentage of difficult variants of the coronary arteries courses (63.2% of cases), which increases the risk of cardiac surgery in early neonatal period. Immediate and long-term results of the two-stage surgical treatment of Taussig-Bing anomaly in combination with coarctation or interruption of the aortic arch demonstrated the safety of this method of surgery (early postoperative mortality and reinterventions were 0%) and low percentage of reoperations (12.5%) in the long-term period.

**Keywords:** Taussig-Bing anomaly, perinatal management, surgical treatment.

**Introduction.** Taussig-Bing anomaly is a variant of double outlet right ventricle (DORV). For the first time this cardiac anomaly was described by Taussig and Bing in 1949 [1]. Later, Stellin et al. described it as a spectrum of anomalies characterized by DORVs, the presence or absence of subpulmonary conus with subpulmonary ventricular septum defect (VSD), the aorta arising completely from the right ventricle (RV), and at least 50% of the pulmonary artery (PA) arising from the RV with parallel (transpositional) course of the great arteries and malalignment of the infundibular septum leading to preferential flow from the left ventricle (LV) to the PA [2]. Hemodynamically, this cardiac anomaly is similar to transposition of the great arteries (TGA) with VSD, requiring appropriate surgical treatment after birth - arterial switch operation with VSD plasty [3]. Likewise, prenatal diagnosis of Taussig-Bing anomaly is similar to diagnostic features of the TGA with VSD [4].

**The aim** of this study was to analyze the features of prenatal diagnosis, perinatal management and surgical treatment in the patients with Taussig-Bing anomaly.

**Material and methods.** During the period from 2006 to 2015 in Ukrainian Children’s Cardiac Center (UCCC) 24 fetuses with Taussig-Bing anomaly were diagnosed during prenatal (fetal) echocardiography. When the diagnosis of anomaly was made before 34 weeks of gestation (w. g.), the fetus was re-examined in 36 w. g. According to our perinatal management all deliveries were planned in Kyiv near the cardiac center with consultation of a newborn in the first hours of life. Echocardiography was performed for all neonates to verify the diagnosis for choice of surgical treatment.

**Results.** This anomaly was diagnosed prenatally by an average age of 27 w. g. (8 cases - up to 22 weeks). The average age of pregnant women at the time of initial consultation was 28,0 ± 5,9 years. In three (12.5%) cases parents decided to terminate a pregnancy. In 19 (90.5%) cases of 21 children were born in Kyiv near the cardiac center. In 2 (9.5%) other cases children were born by place of residence in a specialized maternity hospital: in one case because of the parents’ desire, in the second one because of prematurity (35 w. g.). Among the children born in Kyiv, 17 (89.5%) of 19 were consulted in the first hours of life in the UCCC. In 1 (4.2%) case preterm delivery was occurred with intrapartum death of the child. In the second case, the child was consulted and operated in other cardiac center. In all cases diagnosis of Taussig-Bing anomaly was confirmed. Among concomitant cardiac anomalies, pathology of the aortic arch was dominant – 12 (57.1%) cases (10 coarctations of the aorta, and 2 interrupted aortic arch, type A). Moreover, it was prenatally diagnosed only in 6 (50.0 %) fetuses. In addition, in one (4.8%) case multiple VSDs and in another (4.8%) case atrial septal defect were diagnosed.

Among the variants of coronary anatomy it is worth noting the low percentage of so-called "normal" coronary artery pattern, where left coronary artery (LCA) originates from the left-facing sinus (sinus 1) and right coronary artery (RCA) from the right-facing sinus (sinus 2) (7 cases out of 19, or 36.8%), which is ususal dominant pattern in patients with TGA (about 65%) [5]. Among other options two coronary patterns were dominant: I - left anterior descending artery originated from sinus №1 and RCA with circumflex artery (so-called posterior loop) from sinus 2 (6 cases, or 31.6%); II – RCA originated from sinus 1 and LCA from sinus 2 (so-called double loop) (4 cases, or 21.1%). In addition, in 1 case it was single coronary artery from sinus 1, and in another – two coronary arteries originated from sinus 2 with high take off and intramural course of both arteries.

By 2009, the prostaglandin E1 infusion administered to support the patent ductus arteriosus and Rashkind procedure performed in all patients (4 cases, 100%) with Taussig-Bing anomaly after the birth. Since September 2009, we started the program of surgical correction of TGA in the first hours of life using autologous umbilical cord blood [6]. Therefore, prostaglandin E1 infusion and Rashkind procedure was performed only in patients with Taussig-Bing anomaly when 2-stage surgery (pulmonary artery banding with or without aortic arch angioplasty in the first stage and arterial switch operation with VSD plasty and PA debanding in the second stage) was planned in the future or primary total repair was postponed. In this group of infants, prostaglandin E1 infusion was used in 8 (53.3%) out of 15 cases, and the Rashkind procedure – in 10 (66.7%) cases.

The choice of surgical treatment for the newborns with this pathology depended on the presence of concomitant cardiac anomalies (aortic arch pathology, VSD diameter or multiple VSDs, coronary pattern), gestational age and birth weight. All patients with Taussig-Bing anomaly without concomitant cardiac disease (5 cases), except two cases of premature birth, was made the primary total correction in the average age of 22,4 ± 25,7 days after birth. Two premature babies after Rashkind procedure was performed pulmonary artery banding, respectively on 6 and 76 days of life. In the presence of concomitant aortic arch pathology it were performed total repair in 6 (50.0%) cases on average age of 4,2 ± 4,5 days after birth, and aortic arch angioplasty with pulmonary artery banding in other 6 (50.0%) ones on average age of 2,3 ± 0,8 days of life. In 2-stage surgical treatment of this anomaly, second stage (total repair) was performed after an average of 3-6 months.

In 1-stage (primary) surgical treatment (11 cases), it was 1 (9.1%) intraoperative death through a complex coronary pattern with both intramural coronary course from one coronary sinus. In 2-stage correction (8 cases) it was 1 (12.5%) death of a premature baby in 1 month after the total repair with a history of superior vena cava thrombosis. Postoperatively, 4 re-interventions were after total cardiac repair. Three (27.3%) of them have occurred by 1.5-2.5 months after the primary correction of this anomaly with coarctation of the aorta in the first days of life. These were two endovascular procedures (balloon aortic arch angioplasty because of re-coarctation and balloon pulmonary valvuloplasty due to stenosis) and one reoperation due to obstruction of the LV and RV outflow tracts and right pulmonary artery stenosis. One (12.5%) reoperation after 2-stage way of repair occurred in a patient aged 2 years and 1 month due to obstruction of the RV outflow tract.

**Conclusions.** Prenatal diagnosis of Taussig-Bing anomaly allows scheduling delivery near cardiac center with consultation of a newborn in the first hours of life to determine the optimal management of surgical correction. Particular attention should be paid to the prenatal diagnosis of the aortic arch pathology, which is often associated with this cardiac anomaly and influences the choice of postnatal treatment. In postnatal period an important part of diagnosis is the coronary artery anatomy, as a high percentage of complex coronary patterns increase the risk of primary total repair in early neonatal period. When combined Taussig-Bing anomaly with coarctation or interrupted aortic arch, 2-stage surgery is a safe treatment with satisfactory immediate and long-term results.

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