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The Possibility of Prenatal Echocardiography in The Diagnosis of Agenesis of The Valve of The Pulmonary Artery and Perinatal Outcomes.

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ABSTRACT

Was carried out a retrospective analysis of 10 cases of prenatally diagnosed agenesis of the valve of the pulmonary artery (AVPA). In 100% of cases AVPA in the prenatal period was characterized by expansion of the pulmonary artery and its branches with bi-directional blood flow through the pulmonary valve. When studying four-chamber section a heart cardiomegaly was detected in 3 (30%) cases. The axis of the heart was changed in 9 (90%) cases due to the prevalence of the size of the right ventricle. Agenesis of the ductus arteriosus was detected in 5 (50%) fetuses. The left aortic arch was registered in 7 (70%) cases; right aortic arch – in 3 (30%). The nuchal extension was detected in 2 (20%) fetuses in the first trimester of pregnancy. The term of prenatal diagnosis of AVPA was ranged from 12 to 33 weeks and in the average was 24.4 weeks. Cardiac defects were registered in 5 (50%) cases and extracardiac defects in 2 (20%) fetuses. 5 (50%) cases were involved in the interruption of pregnancy. Three of 5 infants (30%) died, in one case of them after a surgical treatment. Two newborns are alive.

Keywords: fetus, agenesis of the valve of the pulmonary artery, prenatal diagnosis, perinatal outcomes.

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INTRODUCTION

An absence (agenesis) of the pulmonary trunk valve is a congenital heart defect in which the valve of a pulmonary trunk is missing because of the underdevelopment of its leaflets in the period of embryogenesis. Depending on the morphology of the rudimentary valve, there are three forms of defects: 1st form (51,1%) – instead of the valves there is a small circular located fibrous cushion; the 2nd form (32,1%) – the undeveloped leaflets are presented with the small fibrous tubercles; 3rd form (16,8%) – leaflets of valve are available, but they are hypoplastic and sinuses are not formed (Abramian et al., 2011; Wertaschnigg et al., 2013; Malev et al., 2012; Maleva and others, 2013). The frequency of occurrence is 0.2% of all congenital heart defects in newborns. Were described two options of agenesis of the valve of the pulmonary artery (AVPA) that are detected prenatally: more frequent – the first (93%), it is characterized by the absence of the valve of the pulmonary artery with ventricular septal defect (VSD) and agenesis of the ductus arteriosus (tetralogy of Fallot); the second, a more rare variant with an intact atrial septum, with a small degree of enlargement of pulmonary artery and arterial duct (Abuhamad, Chaoui, 2010; Szwast et al., 2014; Volpe, Paladini, Marasini et al., 2004; Sal'nikov, 2000; Sal'nikov et al., 2012; Sal'nikov, Lobanov, Davtyan, 2009; Komarova, Ivanova, 2013).

A ring of the pulmonary trunk valve can be widened, narrowed or hypoplastic. This defect is isolated very rarely, usually AVPA is associated with a stenosis of infundibulum of the right ventricle and with VSD, and the internal structure of the heart resembles the tetralogy of Fallot. In the first variant of the defect (like Fallo) the risk of chromosomal abnormality is high enough. In 40% of AVPA cases is detected a microdeletion 22q11, and are also described cases of trisomy 13 and 18. Therefore, the identification of AVPA like Fallo in the fetus dictates the necessity of prenatal karyotyping and FISH-diagnosis. In the second variant of AVPA with intact atrial septum the chromosomal and extracardiac anomalies are rarely recorded.

Publications on prenatal ultrasound diagnosis of AVPA are mainly represented only by a few clinical observations as in the Russian (Galindo et al., 2006; Burakovsky, Bockeria, 1989; Martynov et al., 1995; Medvedev, 2012; Onischenko, 2005; Potolova, Kopylova, 2013) and foreign (Gereshti, 2012; Rybakova, Alekhin, Mitkov, 2008; Tkachenko, Beresten, 2006) periodicals. Therefore, the aim of our study was a generalized analysis of 10 cases of prenatal diagnosis of AVPA.

METHOD

In the analysis were included 10 observations of AVPA. At ultrasonography all the patients underwent an extended echocardiography and a detailed anatomical study of the fetus to exclude concomitant pathology. For the analysis were chosen the following variables: maternal age, gestational period of establishment of prenatal diagnosis, the presence of the ductus arteriosus, associated cardiac and noncardiac anomalies.

In the study of the fetal heart were included the following criteria: four-chamber section of the heart with the assessment of axis of the heart, section through the three vessels and trachea, a section through the output tracts, the location of thoracic aorta and blood flow parameters in the pulmonary artery.

The age of pregnant women was ranged from 25 to 32 years and the average was 28.5 years. There were 2 women who were pregnant in a first time (20%), those who were pregnant before – 8 (80%). Pregnancy with one fetus was diagnosed in 9 cases and in one case – monochorionic twins. Weight of newborns was from 3400 to 3590 g, and in an average - 3483 g.

An assessment of the location of the axis of the fetal heart was carried out in a relation to the sagittal plane in the study of the four-chambered heart. The normative values of the angle between the axis of the fetal heart, passing through the interventricular septum, and a sagittal direction in the second half of pregnancy were considered from 30° to 60° [2].

An assessment of the location of the thoracic descending aorta was performed in the study of four-chamber section of the heart. A normal cross-section of thoracic aorta of the fetus is placed to the left of the sagittal plane [4].

Cardiothoracal correlation (CTC) was defined in terms of four-chamber fetal heart. The width of the heart was measured at the level of the atrioventricular valves, the chest – on the outer contour of the ribs. The CTC that was more than 50% was considered as cardiomegaly.

RESULTS

Diagnosis of AVPA was established in the first trimester in one case (10%), in the II trimester - in 6 (60%) and in the III trimester – in 3 (30%). The average time of diagnosis of AVPA was 24.4 weeks. Before 22 weeks were diagnosed 4 (40%) cases of AVPA. In 2 of these 4 cases AVPA was associated with cardiac pathology, in one with right aortic arch. In 5 (50%) cases of AVPA was marked the agenesis of the ductus arteriosus. The average time of diagnosis of AVPA in this group was 29 weeks.

When studying four-chamber section of the heart a cardiomegaly was revealed in 2 (20%) cases in the second trimester of pregnancy.

In one case AVPA was combined with a stenosis of two leaflets of valve of the pulmonary artery, in the other – with Uhl anomaly, dysplasia of the tricuspid valve and the diverticulum of the right ventricle. It can be concluded that cardiomegaly (CTC of more than 50%) is found in fetuses, when AVPA is combined with the other cardiac pathology in the second trimester of pregnancy. In one case, cardiomegaly was detected in the first trimester of pregnancy in fetuses with AVPA and its origin is associated with the volume overload of the ventricles of the heart.

The axis of the heart was changed in 9 (90%) cases, median amounted to 81,40°. The change in the heart axis was mainly attributable to the predominance in size of the right ventricle over the left.

An abnormal position of the thoracic aorta was recorded in 3 (30%) cases that were accompanied in all cases with a right aortic arch. In 2 cases the thoracic aorta was located to the right, in one – in the central. In all three cases the AVPA was combined with right aortic arch and the agenesis of the ductus arteriosus.

In the section via the output tract of the right ventricle in all 10 cases (100%) was marked an increase in the pulmonary artery and its branches.

The diameter of the pulmonary artery exceeded the 95th percentile of normative performance. With the use of color Doppler mapping in all cases were registered the bidirectional blood flow in the pulmonary artery.

In the section via the output tract of the left ventricle in 9 cases was discovered the presence of an output of the ventricular septal defect with a displacement with/without expansion of the aortic root.
In 1 case the interventricular septum was intact and this case was highlighted as AVPA of the II type. It was associated with cardiac pathology - an Uhl anomaly.

DISCUSSION

The most revealing prenatal ultrasound marker of AVPA is a section through three vessels and trachea, and an output portion of the right ventricle, where in all studied fetuses, regardless of gestational age, was marked the 100% extension of the main trunk of the pulmonary artery with branches. When using a color Doppler mapping in the pulmonary artery were recorded an antegrade blood flow in the phase of systole and a retrograde flow in the phase of diastole.

Among the concomitant heart diseases were registered: dysplasia of tricuspid valve in 2 cases, double outlet of right ventricle (1), Uhl anomaly (1), ventricular diverticula (2), aortic valve stenosis (1), bicuspid aortic valve (2), the left aberrant subclavian artery (1), right aortic arch (3). Extracardiac pathology was found in two cases: agenesis of the kidneys and hypoplasia of the thymus (20%).

The perinatal outcomes of prenatal diagnosed cases of AVPA in our analysis were as follows: in 5 (50%) cases the pregnancy was terminated, 5 (50%) children were born alive in time. Among them in 2 cases

was occurred a neonatal death, one child died after the operation and only two children survived. One was operated; the second is waiting for the operation. Perinatal loss was 80%.

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Thus, a screening ultrasound examination in the II and the III trimester of pregnancy with the study of four-chamber sections of the heart, section through the three vessel and sections across the output tracts of the ventricles, allows making a prenatal diagnose of AVPA in 100% of cases. The most typical symptoms of AVPA are the extension of the main trunk of the pulmonary artery and its branches, and bi-directional blood flow in the pulmonary artery. A perinatal loss was 80%, which indicates a poor prognosis in fetuses with a prenatally diagnosed AVPA.

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