Original **paper**

SINGLE FETAL CARDIAC TUMORS AND FOLLOW-UP BASED ON 13 CASES FROM THE FETAL CARDIAC REFERRAL CENTER IN 1993-2017

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Abstract

Introduction: Fetal cardiac tumors are anomalies, that occur rarely: from Nationwide Register of Fetal Cardiological Problems in Poland in years 2004-2016 amongst 8112 fetuses with cardiological problems, there were 85 fetuses with cardiac tumors, including 52 cases of multiple cardiac tumors (0,64%) and 33 of single anomalies (0,4%).

Material: This analysis included 13 cases from single tertiary fetal cardiac center Lodz in years 1993-2017.

Results: Ten out of 13 fetuses with single cardiac tumors (SFCT) had cardiomegaly: on average HA/CA was 0,49. The size of the tumor was different: the smallest one – 6 x 6 mm, the biggest 47 x 47 mm. The way of the delivery: in 10 cases there was CS and in 3 cases natural delivery. Birth weight was from 2000-3950 g (average 2989,2 g). Cardiosurgical resection of the tumor was performed on 4 newborns: at 2nd, 4th, 8th and 16th day of life (average 7,5 day). Four neonatal deaths were registered (31%): in 1st 2nd and 11th day (before surgery) and in the 28th day after the operation.

Conclusions: Single fetal cardiac tumors (SFCT) can be diagnosed at 20 weeks of pregnancy, which allows to start echocardiographic monitoring, taking into consideration the potential risk of hemodynamic progression. SFCT can be the first sign of tuberous sclerosis complex in later prenatal or postnatal life. SFCT other than rhabdomyoma can be asymptomatic in newborn, but may require an early cardiosurgical resection.

Key words: *congenital heart disease, fetal echocardiography, cardiac tumor, prenatal diagnosis*

INTRODUCTION

Fetal cardiac tumors are rare anomalies: 0,08-0,2%[1,2,] while data from Polish Nationwide Register of Fetal Cardiological Problems (in polish- ORPKP), suggested higher prevalence (1,05%). In years 2004-2016 amongst 8112 fetuses there were 85 fetuses with cardiac tumors, including 52 cases of multiple cardiac tumors (0,64%) and 33 of single anomalies (0,4 %) (Chart 1).

According to the literature, most of the fetal cardiac tumors are primary anomalies, and dominating patomorphology type is multiple rhabdomyoma (60%-90%), very well described and the proceeding during prenatal time and after delivery is commonly known[2,3,4].

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Single fetal cardiac tumors are some sort of enigma and for now, there is no unitary algorythm of proceeding, that is why it become the aim of our work.

MATERIAL

The analysis included 13 cases from our unit Fetal Cardiology Department - Referral Center in the middle of Poland, during the years 1993-2017. Study group were women at the age of 18-37 years, average gestational age during targeted echocardiography examination was 34 weeks of pregnancy. The earliest registration of cardiac tumor was in 20th week of pregnancy, and the latest was during perinatal period (40th week of pregnancy).

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Clinical data on the analyzed material are presented in Table 1, including age of pregnant woman, gestational age of the fetus during echocardiography examination, way of delivery, neonatal birth weight, information of cardiosurgical treatment and histopathological diagnoses.

Data concerning the results of the echocardiography examination are presented in Table 2 and Chart 2. Single cardiac tumors were most often accompanied by cardiomegaly (10/13), pericardial effusion (6/10), tricuspid regurgitation (3/10) and myocardium hypertrophy (3). Other functional echocardiographic anomalies such as aortic valve stenosis, foramen ovale restriction, right atrium enlargement, arrhythmia (premature atrial contractions), mitral valve stenosis and pulmonary insufficieny were present in two or single fetuses.

Data concerning extracardiac anomalies are presented on Table 3 and Chart 3: in two fetuses elevated amount of the amniotic fluid was diagnosed and in 2 decreased amount (one was diagnosed with oligohydramnion). Single fetuses were diagnosed with CNS tumor, kidney tumor, hydrothorax, unilateral pyelectasis, hydrops testis and situs inversus (stomach on the right side). One fetus presented small gestational age with normal Doppler blood flow in umbilcial cord and middle cerebral artery.

Follow-up of the fetuses and newborns are presented on Illustration 1, including cardiosurgical operations or discharge from the hospital without surgical intervention.

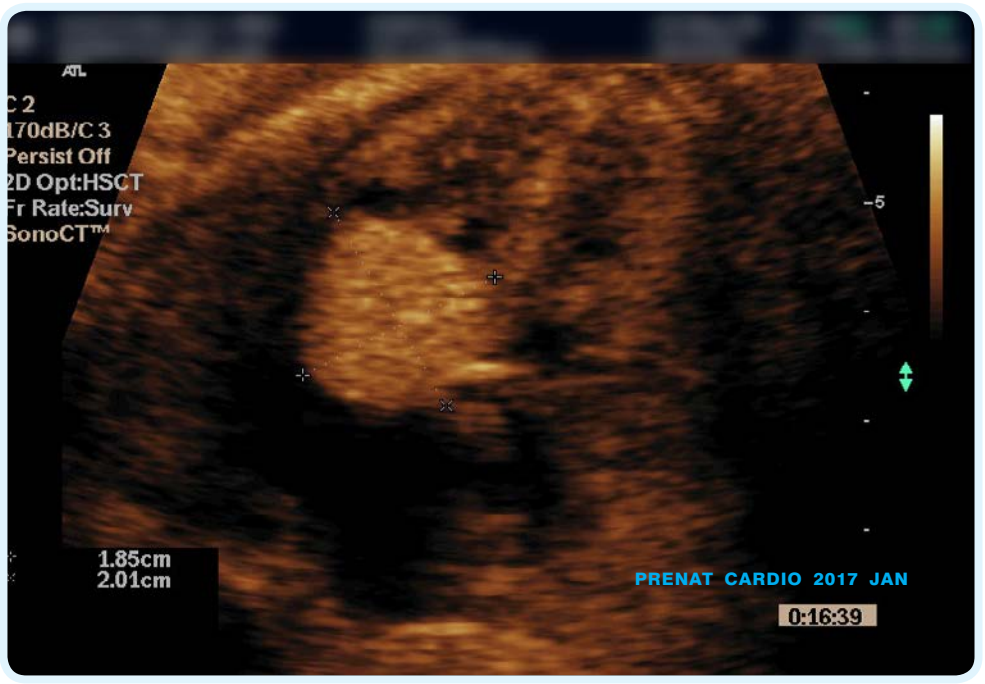
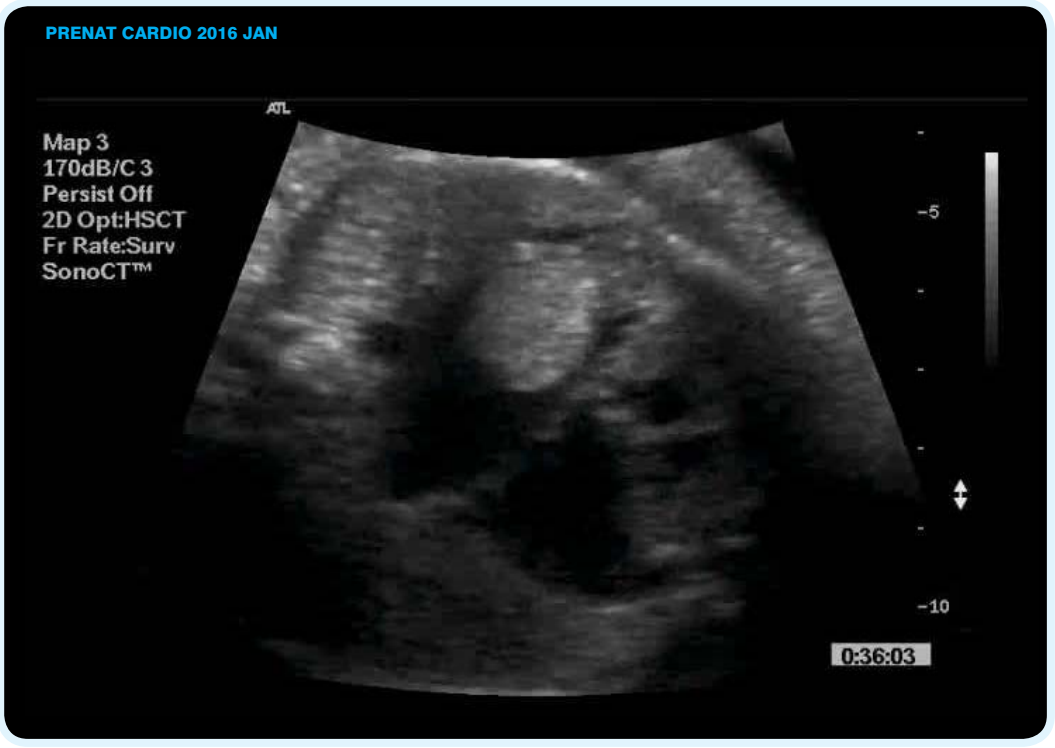


Fig.1-1. Fibroma.

RESULTS:

In the group of 12 fetuses, normal prenatal development was observed, which means that fetal age according to the last menstrual date was correct in comparison to the biometric age. One fetus (position 1 in Table 1), was small for gestational age, however Doppler blood flows in umbilical cord and middle cerebral artery were normal.

In 9 fetuses the amniotic fluid index (according to Phelan method) was correct, in 2 there



Fig, 1-2 (Clne): Fibroma.

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was elevated AFI up to 28, in 1 fetus there was oligohydramnion AFI 4 cm and in 1 with AFI 7,5 observation towards oligohydramnion was suggested.

Extrcardiac anomalies: in one case, in CNS a tumor in the front corner of the lateral right ventricle was observed with symetrical dilatation of lateral brain ventricles , at the height of the posterior corners, up to 21 mm, in another case in the male fetus unilateral hydronephrosis up to 7,5 mm was present. In the single fetus there was situs inversus.

In 3 fetuses functional abnormalities were observed: hydrothorax (1x), pyelectasis (1x) and hydrops testis (1x) (Table 3, Chart 3).

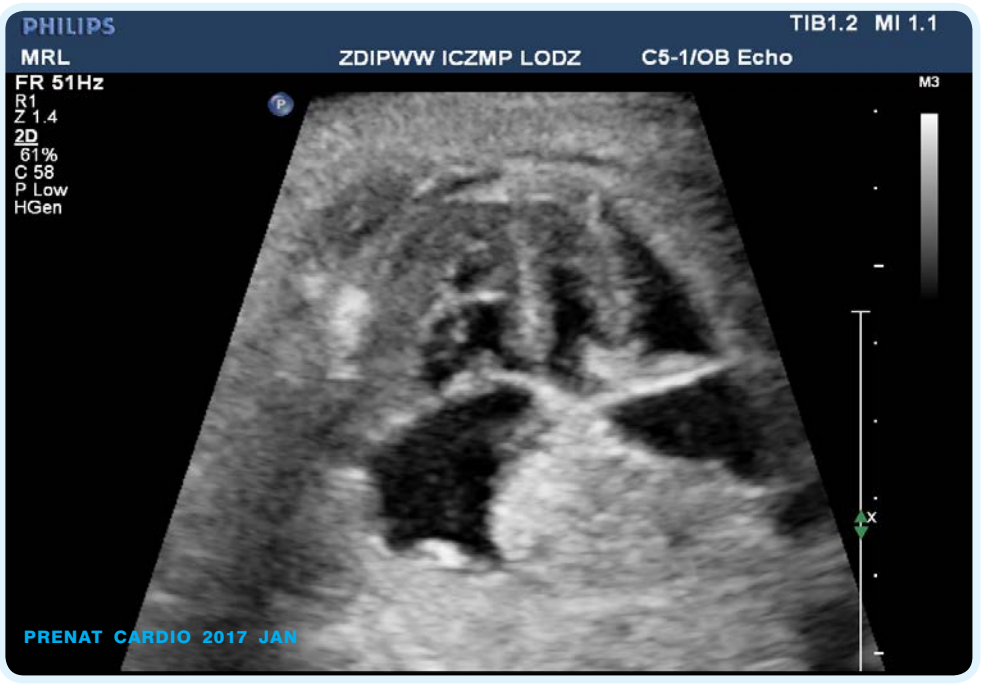
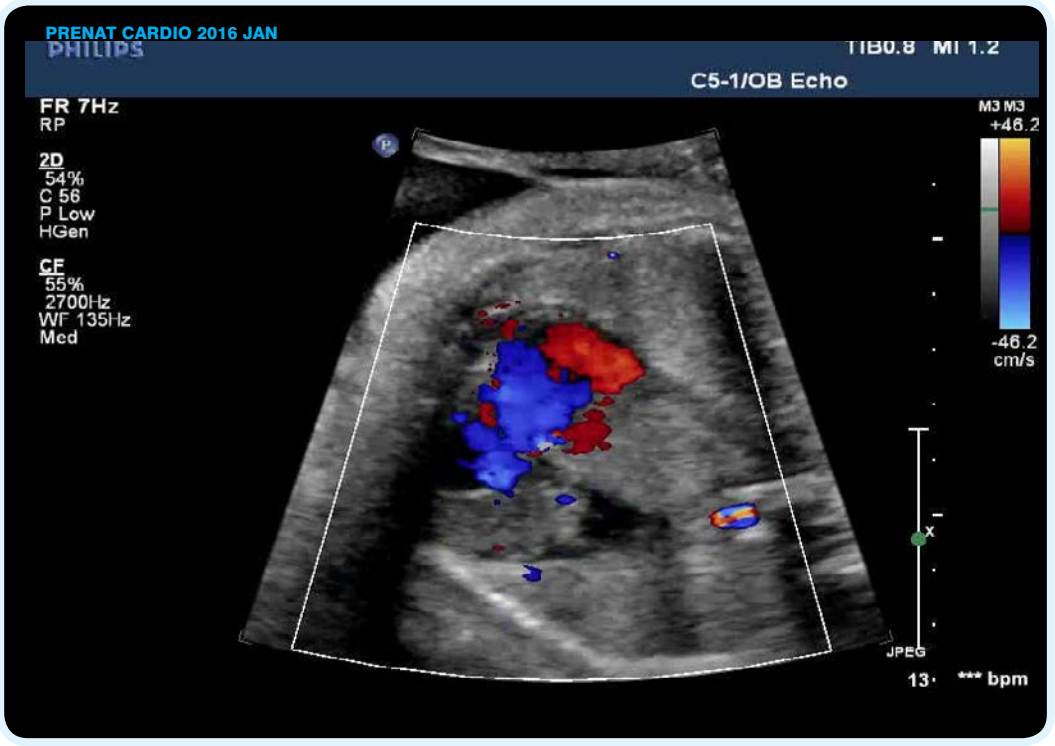


Fig. 2-1. Teratoma

In 10/13 fetuses with single cardiac tumors, cardiomegaly occured: on average HA/CA was 0,49. In 7 fetuses a pericardial effusion >3mm was present, in 3 fetuses there was tricuspid insufficiency also in 3 fetuses myocardial hypertrophy occured. In one case, in the course of the single tumor mass (case nr 7) there was an increased blood flow through the aortic valve, up to 150cm/s and also reverse flow in the hypoplastic ascending aorta. In 1 fetus, at 28th week of pregnancy, restrictive flow through foramen ovale was observed, FO was 2 mm. In single fetuses there was enlargement of the right atrium, in another case arrhythmia ocured – supraventricular extra beats, in one fetus was mitral insufficiency and also in one case pulmonary insufficiency.

The size of the single tumors was different: the smallest one – 6 x 6 mm, the biggest 47 x 47 mm.

The way of the delivery: in 10 cases there were cesarian sections, in 3 cases natural delivery. Birth weight was from 2000-3950g (average 2989g). Newborns gained on average 7 points in Apgar scale. Time of the hospitalization of the newborns was from 5 to 53 day (without one-day hospitalization of the newborn that passed away in his first day of life. Cardiosurgical resection of the tumors was performed on 4 newborns: at 2nd, 4th, 8th and 16th day of life (average 7,5 day).



Fig, 2-2 (Clne): Teratoma

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There were 4 deaths (31%): in 1st, 2nd and 11th day (before the resection of the tumor) and in the 28th day after the cardiosurgical operation.

Nine patients were discharged in general good health, without clinical symptoms, for further observation, witout the need of cardiosurgery.

Nr	AGE	HRP	HBD	SEX (FETUS)	DELIVERY	BIRTH WEIGHT (g)	SURGERY	DAY OF SURGERY	DEATH	RESULT
1.	37	1	38	F	CS	2000	NO		YES	R
2.	22	1	35	F	CS	2800	NO		YES	R
3.	37	1	31	F	CS	3200	YES	8	NO	R
4.	22	0	37	M	CS	3300	YES	2	YES	F
5.	30	0	35	M	V	2900	YES	16	NO	T
6.	33	0	26	M	V	3700	NO		NO	UK
7.	24	1	34	M	CS	2950	NO		NO	UK
8.	36	1	28	F	CS	2400	NO		YES	UK
9.	29	1	40	M	CS	3950	NO		NO	UK
10.	34	1	28	M	CS	2220	NO		NO	UK
11.	22	1	20	M	CS	3250	NO		NO	UK
12.	18	0	31	M	V	3000	NO		NO	UK
13.	31	0	31	M	CS	3190	YES	4	NO	T

Legend: RESULT - Result of histopathological examination /// HRP – high risk of pregnancy /// T – time – time of detection the abnormality in the heart in Hbd /// M – male sex /// F – female sex /// CS – delivery via c-section /// V – vaginal delivery /// R – rhabdomyoma /// F – fibroma /// T – teratoma /// UK – unknown, lack of information about histopathology

Table 1. The table presents data of examined population (ICZMP Prenatal Cardiology Centre 1993-2017)

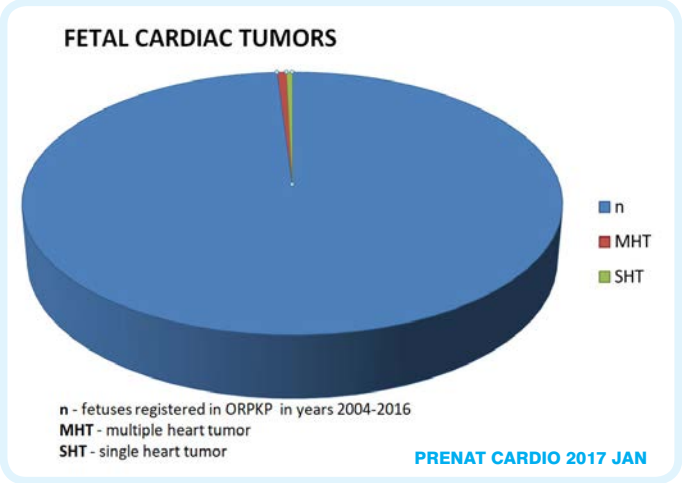

Chart 1. According to the data from National Registry in Poland ORPKP during years 2004-2016 amongst 8112 feuses with cardiac problems, 85 fetuses were diagnosed with 52 cases of multiple cardiac tumors and 33 cases with single cardiac tumors, (in total 1,05%).

DISCUSSION:

Single tumors has no predilection towards localization in specified heart cavity[5, 6]. The analysis from our Center revealed, that tumors were located both in the atrium, heart ventricles or intraventricular septum. The dimensions of the abnormalities were different, from a few milimeters up to few centimeters. In most cases, presence of the single fetal cardiac tumor had no influence on the fetuses growth, which went on without problems (n=12).

Early prenatal detection of the single tumor mass, within the heart of the fetus, demanded long-term echocardiographic monitoring, to keep the pregnancy safe[7]. In most cases, the pregnancy could be continued until the due day and there was no need of delivering before the deadline.

In differential diagnosis, hyperechogenic spots

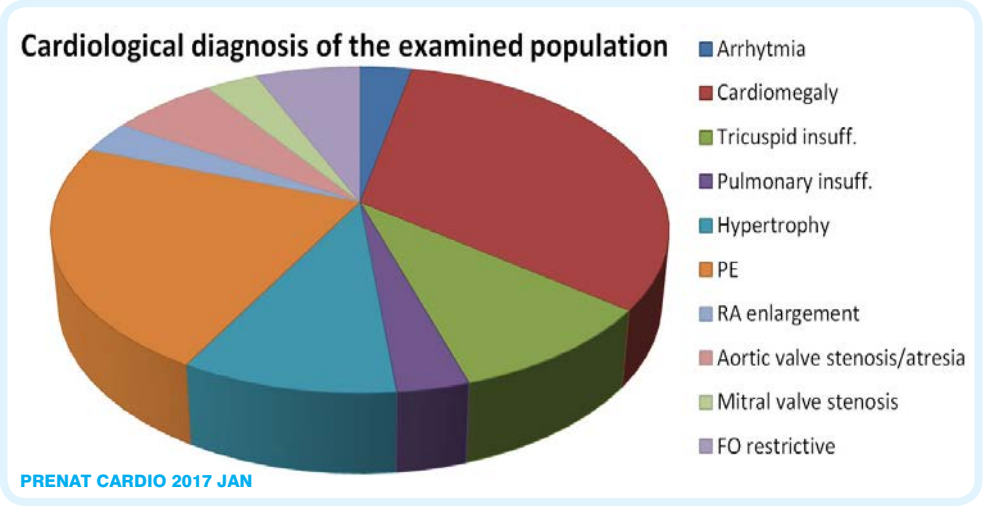

Chart 2. Data from ICZMP Prenatal Cardiological Centre (1993-2017) in 13 fetuses with single heart tumor.

Cardiological diagnosis of the examined cases (1993-2017)	Number of fetuses out of 13
Cardiomegaly	10
Pericardial Effusion	7
Tricuspid insuficiency	3
Myocardial Hypertrophy	3
Aortic valve stenosis	1
FO restriction	1
Right Atrium enlargement	1
Arrhythmia	1
Mitral valve insufficiency	1
Pulmonary insuff.	1

Table 2.Data from ICZMP Prenatal Cardiological Centre (1993-2017). Register of cardiological diagnoses amongst examined population.

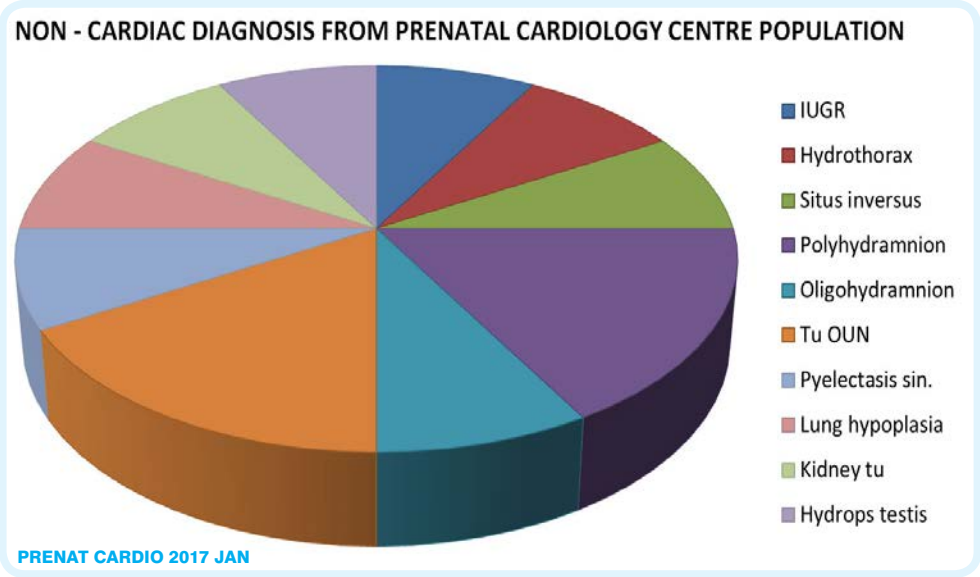

Chart 3. Non-cardiac diagnosis from ICZMP Prenatal Cardiology Centre population (1993-2017)

Non-cardiac diagnosis of the examined cases(1993-2017)	Number of fetuses
Polyhydramnion	2*
Oligohydramnion	2**
CNS tumor	1
Kidney tumor	1
Hydrothorax	1
Pyelectasis sin.	1
Situs inversus	1
Small gestational age	1
Hydrops testis	1

**AFI (amniotic fluid index) – a sum of 4 fluid spaces in the pregnant uterus, in case of patient with polyhydramnion AFI – 28*
***Oligohydramnion AFI – 4 cm (one fetus) and AFI 7,5 cm (other fetus - observation towards oligohydramnion)*

Table 3. Extracardiac anomalies in 13 fetuses with single heart tumor (Prenatal Cardiology Dept 1993-2017)

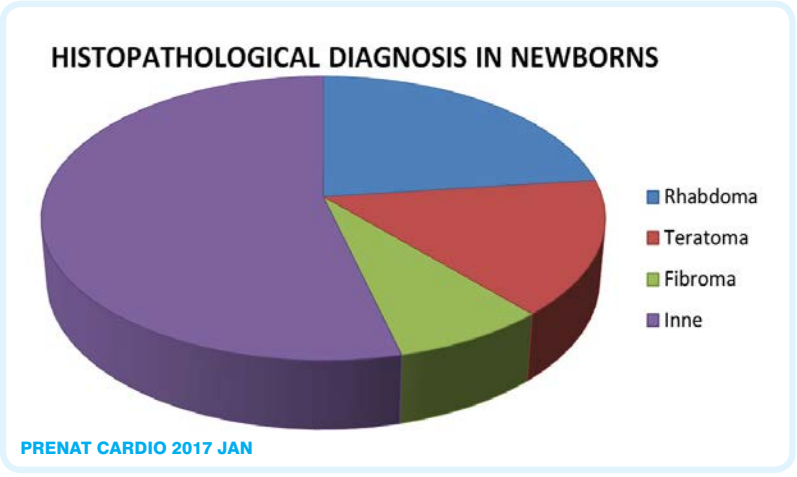

Chart 4. Histopathological diagnoses in 6 newborns: 4 had cardiosurgical operation (successful resection in 3, death of 1 newborn) and 2 deaths before surgery

chambers, known as "bright spots", which can be misinterpreted as small cardiac tumors, were taken into concern. In US examination these are homogenous, hyperechogenic nodule with benign contours and oval shape, known in the literature also as "golf ball sign". Because of possibility of the progression, first known as "bright spot" and finally diagnosis as a cardiac tumor, intense observation in selected cases is justified, however in our analysis there were no fetuses with primary classified abnormality as "bright spot".

Socolowski[8], in his publication concerning EIF (echogenic intracardiac focus) in patients with prenatal diagnosed hyperechogenic spot in heart cavity, pointed out the possible role of the infectious factor, which can also be the cause of low birth weight in these cases.

In fetuses, according to prenatal ultrasonographic-echocardiographical diagnostic, there is need to make an attempt to determine the prognosis, even without the lack of information about histopathology, which can be obtained only after the tumor resection, usually during neonatal period That is why attempts of determing the prognosis at the prenatal stage of patients life are much more difficult than in the case of pediatric cardiology.

In our material, in subgroup (n=6) with histopathology exams, the most common single cardiac tumor was rhabdomyoma (3x), followed by teratoma 2x and in one case there was fibroma, (Chart 4). In recent literature, for the first time, we pay attention to unusual course of "rhabdomyoma cordis", which can manifest itself not only as a multiple tumor but as a single cardiac tumor, without any additional changes in CNS or kidneys. In one case of single cardiac tumor, presence of additional tumor in CNS suggested suspicion of rhabdomyoma during prental life, which was confirmed later on during boy's treatment in our hospital (Diagnosis "SEGA"). In the "classic" way, according to the literaure, tuberous sclerosis complex usually reveals itself in the 1-2 year of life, in child that was earlier observed because of multiple fetal cardiac tumors[3,4].

In echocardiographic examination cardiac tumors were morphologically homogenous, hyperechogenic with benign and regular contours and oval shape, likewise in Carvalho et al publication[9]. Unregular shape and echogenicity occured in our series, in teratoma tumor case (patient nr 5, Table 1).

Fibroma is much rarer histopathological variety (6-25% according to the pediatric cardiology) with prediction to invasing interventricular septum or left heart ventricle. In our series, we observed 1 case (patient nr 4, table 1). Usually, it is relatively large mass, with tendency to progress in contrast to rhabdomyoma, with tendency to regress after delivery. According to the literature [10] fibroma occurs relatively more often than rhabdomyoma with fetal heart arrhythmia, but our observations do not confirm this. In most patients from literature, with diagnosed fibroma, sudden death was observed, due to hemodynamic disorders, that were caused by disturbed blood flow in artio-ventricular connections or blood flow in vessels coming from both ventricles. The stage of heart failure was correlated with size and localization of the tumor[10].

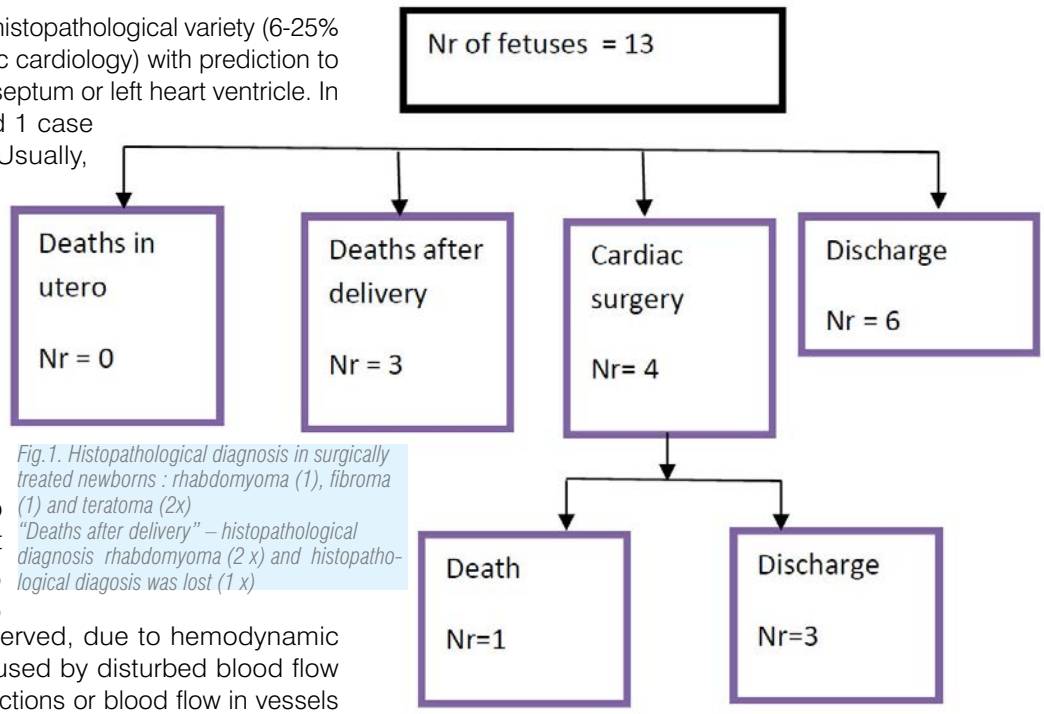

Fig.1. Histopathological diagnosis in surgically treated newborns : rhabdomyoma (1), fibroma (1) and teratoma (2x)
"Deaths after delivery" – histopathological diagnosis rhabdomyoma (2 x) and histopathological diagosis was lost (1 x)

In comparison to others, teratoma occurs not in the heart, but with close contact with aorta or pulmonary artery[11, 12]. Fetuses with teratoma are presenting various clinical symptoms, from asymptomatic, to mild pericadial effusion up to hydrops fetalis. Yinon et al. reported about 40 cases of fetuses with teratoma and none of them survived [13]. Sydorak et al. described 31 year old patient, at 24 weeks of pregnancy with teratoma and in this case hydrops fetalis occured[11].

There was an attempt to remove a mass from access via chest of the fetus, but the newborn died after the delivery. Rychik described a case of 23 year old pregnant woman, with a fetus diagnosed at 22 weeks with cardiac tumor and in the 24 week of pregnancy the tumor was succefully resected. Newborn was delivered via c-section at 38 weeks of pregnancy and had no cardiac symptoms[14].

In fetus, that we were observing, with cardiac tumor in the form of teratoma (patient nr 5, Table 1), despite the growing of the tumor in the last 3 weeks before the delivery, intercardiac flows were normal, which allowed to continued pregnancy until the due. Clinical condition of the newborn , delivered via c-section was good. He was asymptomatic with no heart murmur. In the 4th day of life, resection of the tumor was made with a good outcome.

Myxoma is another single cardiac tumor, but was not presented in our material. In the literature, Paladini reproted one such case: presented as a right atrial mass, at 23 weeks' gestation. It was followed until delivery and removed at 20 days of life[15].

Zhou et al. reported 14 cases of prenatal diagnosis of hemangioma, mostly located in the right atrium. All fetuses died[16].

Based on our analysis, despite the unfavorable prenatal diagnosis, there was no need of delivering preemies, in most cases, we did not observed significant hemodynamic problems. Because of echocardiographic monitoring, hemodynamic state of the fetuses was controlled and there was a chance to continue the pregnancy safely.

CONCLUSIONS:

Single fetal cardiac tumors can be diagnosed at 20 weeks of pregnancy, which allows to start echocardiographic monitoring, taking into consideration the potential risk of hemodynamic progression.

Single cardiac tumor can be the first sign of tuberous sclerosis complex in later prenatal or postnatal life.

Single cardiac tumor other than rhabdomyoma can be asymptomatic in newborn, but may require an early cardiosurgical resection.

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K. Michalak - work with the manuscript

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E. Czichos - histopathology evaluation, work with the manuscript

H. Romanowicz- histopathology, work with the manuscript

M.Slodki - fetal echocardiographer, work with manuscript

M. Respondek-Liberska - concept of the research, final version

Conflict of interest: The authors declare no conflict of interest