PULMONARY VALVAR STENOSIS IN POLISH NATIONAL REGISTRY FOR FETAL CARDIAC MALFORMATIONS

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Abstract

Prenatal cardiologists for councelling, most often are using both terminology and data from pediatric cardiology, which is a population of different age.

As there is lack in polish literature data about pulmonary stenosis in prenatal population, we retrospectively evaluated our data from National Registry of Cardiac Problems in Fetuses.

Key words: *pulmonary valvar stenosis, congenital heart disease, registry*

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Prenatal cardiologists during consultation of pregnant woman according to heart defects of her fetus most commonly are using terminology and data retardant to pediatric cardiology, patient population in another age. Considering the lack of data in publications on the prevalence of fetal pulmonary valve stenosis in Poland, authors completed a review of the material in the National Register of Cardiovascular Pathology in fetuses.

MATERIALS AND METHODS:

In total, 71 cases was qualified for a retrospective analysis of fetuses with isolated pulmonary valve stenosis (code defects 05.09.04). For this analysis minimal ORPKP base of standard reports was applied, authors presented basic types of heart defects in fetuses regarding to the area of PS, PS position was also compared towards Polish national database to database structure in London by Allan and Sharland.

RESULTS:

Isolated fetal pulmonary valve stenosis is a rare defect, regarding to it's frequency by ORPKP it can be found in position No. 18. More often occurred cases of for example : stenosis, aortic or tricuspid atresia (Table 1). Annually, to this database is introduced 1 to 7 cases of fetuses with isolated heart defect in the form of pulmonary valve stenosis (PS). It has been shown that slight upward trend occurred, especially in the last 4 years (Figure 1). The highest number of PS were from centers LSF PBU n = 21 n = 18 n = 13 YGH, HWQ YBX n = 6 n = 4 (Table 3). Other facilities introduced isolated cases.

In 48/71 cases analyzed pregnancy was nulliparous (67%) and in other 23/71 - multiparous (33%), while in 18 cases it was second pregnancy, four cases of third pregnancy, in one case - the fourth pregnancy. On average, the time needed to detect heart defects was after performed third ultrasound. On average, the diagnosis of heart defects in the form of pulmonary valve stenosis was found in 27 weeks pregnant +/- 7 weeks. The disadvantage of the fetus in the form of PS were recorded starting from 14 to 37 weeks of pregnancy, whereas the detection of growth was observed in the mid-pregnancy between 18 and 22 weeks, but also in the 3rd trimester of pregnancy (Figure 3). In 64 cases there was a natural

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	Types of fetal CHD in Polish Registry	Nr of cases	
1	Hypoplastic left heart syndrome	505	10.2%
2	AVSD: atrial & ventricular components (complete)	411	8.3%
3	VSD	311	6.3%
4	Tetralogy of Fallot	255	5.2%
5	Complete transposition of great arteries (IVS) - proste TGA	202	4.1%
6	Twin-to-twin transfusion syndrome	186	3.8%
7	Aortic valvar stenosis	185	3.7%
8	Double outlet RV - transposition type	139	2.8%
9	Muscular VSD	116	2.3%
10	Perimembranous VSD	115	2.3%
11	Inlet VSD	114	2.3%
12	Supraventricular tachycardia	113	2.3%
13	Discordant VA connections (TGA) w wadach złożonych nie proste TGA	106	2.1%
14	Common arterial trunk	105	2.1%
15	Aortic coarctation	104	2.1%
16	Tricuspid atresia	85	1.7%
17	Double outlet RV - Fallot type	75	1.5%
18	Pulmonary valvar stenosis	71	1.4%

Table 1. Eighteen the most common CHD in Registry (ORPKP) in years 2004-2013

Lp	Type of CHD in London Registry	Nr of cases	100%
1	AVC	375	17,5%
2	HLHS	350	16,4%
3	VSD	200	9,3%
4	CoA	150	7,0%
5	MiA	125	5,8%
6	TriA	80	3,7%
7	KMP	75	3,5%
8	Fallot	70	3,3%
9	Atrezja z płucnej bez VSD	65	3,0%
10	Dysplazja z trójdzelnej	60	2,8%
11	Krytyczna AS	58	2,7%
12	DORV	57	2,7%
13	CoA	54	2,5%
14	Ebstein	50	2,3%
15	TGA	49	2,3%
16	Double Inlet V	28	1,3%
17	PA + VSD	25	1,2%
18	Tumor	25	1,2%
19	Truncus	24	1,1%
20	Pulmonary valvar stenosis	23	1,1%

Table 2. Twenty the most common fetal congenital heart defects in Allan and Shurland (London Registry) data base from 1980-1997 (17 years n=2136)[9]

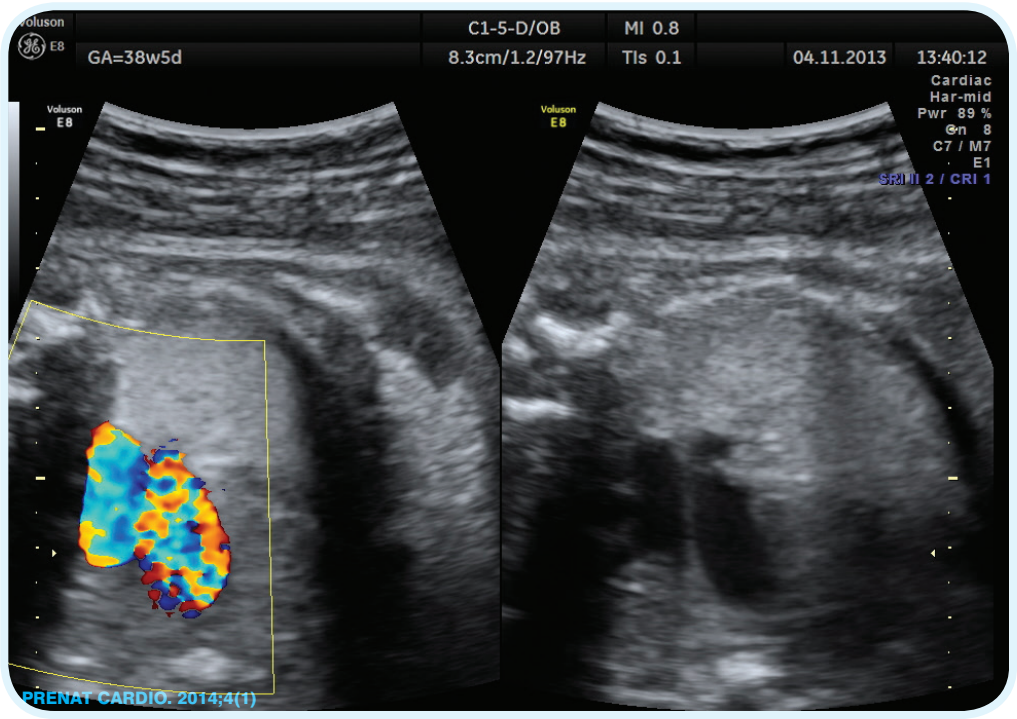

Photo 1. Presentation 2DD + KD and 2D in fetus with critical pulmonary stenosis: in color Doppler blood flow there is turbulent (green-yellow) blood flow and in 2D presentation at the same level there is posstenotic dilatation of pulmonary artery outflow tract. Foto from www.ORPKP.pl by J.Paluszynski MD, PhD from Toruń (code 19861207-ma-dzw-wąb-c1)

fertilization process and in 7 cases, the pregnancy was the result of in vitro fertilization. One case involved a twin pregnancy. 47 pregnancies were low-risk, 24 cases was reported as high-risk pregnancy. Based on 21 completed forms relating to births and the fate of the newborn know that: infants were born with average body weight in 2935 (min 1070 max 4500g). 17% cases of deaths was detected.

DISCUSSION

The incidence of isolated fetal pulmonary valve stenosis (PS) in the existing literature was not precisely described. On the one hand, accompanied by a narrowing of the pulmonary valve in 25-30%

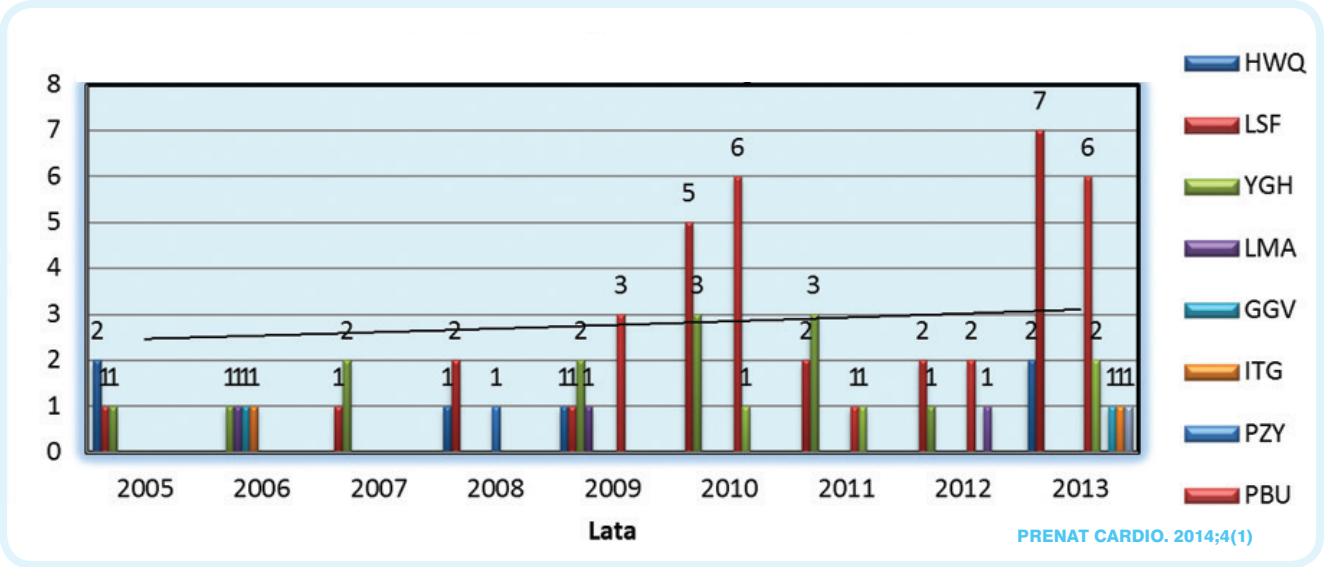

Figure 1. Nr of fetuses with isolated PS in ORPKP.pl in years 2005-2013

of all defects of the heart. On the other hand, the pediatric population incidence of pulmonary valve stenosis is estimated at 8-10% of the total heart defects. If a parent is affected by pulmonary valve stenosis, the likelihood of PS of his child is 2.8%. Regarding to twins PS occurs in 8.3 % cases of monozygotic twins and in 2.2% of those which are dizygotic. Pulmonary valve stenosis occurs as isolated disease, as well as with teams of genetic defects (eg. Z. Noonan, Z. Williams). In the first Polish textbook "fetal echocardiography and cardiology", this defect was not extracting as a separate unit, but it was described in the section on aortic pulmunary atresia. Similarly, in the manual by J. Dangel of the year. 2007. In the manual "Fetal Cardiology" Yagel, Silverman and Gembrucha JC Fouron says that in its database of 40 fetuses 36 had mild (mild-to moderate), manifesting only an acceleration of blood flow through the pulmonary valve> 150 cm / sec (using standard 80 cm / sec). Such anomalies are relatively difficult to detect, because there are 4 cardiac that remains normal, while only during the time of listening intracardiac flow the anomaly can be identified. Newborns have an excellent prognosis and do not require special action after birth. Such defects according to a new division of cardiac defects are not urgent qualify of defects. In contrast, a critical narrowing of the pulmonary valve is a further disadvantage of both the fetus, as well as in newborn. This defect occurs prenatally with a significant wave of tricuspid regurgitation, the light of the right ventricle may be overgrown,

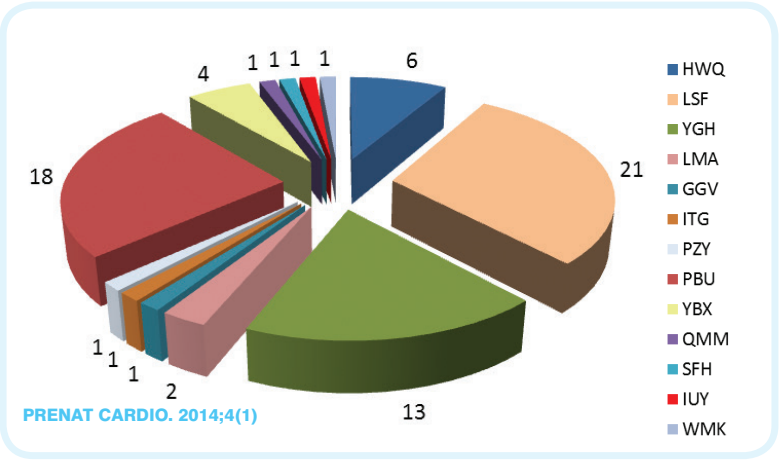

Figure 2. Nr of fetuses with isolated PS in different centers.

	2005	2006	2007	2008	2009	2010	2011	2012	2013	RAZEM
HWQ	2			1	1				2	6
LSF	1		1	2	1	5	2	2	7	21
YGH	1	1	2		2	3	3	1		13
LMA		1			1					2
GGV		1								1
ITG		1								1
PZY				1						1
PBU					3	6	1	2	6	18
YBX						1	1		2	4
QMM								1		1
SFH									1	1
IUY									1	1
WMK									1	1

Table 3. Nr of fetuses with isolated PS in years 2005 2013 in different centers

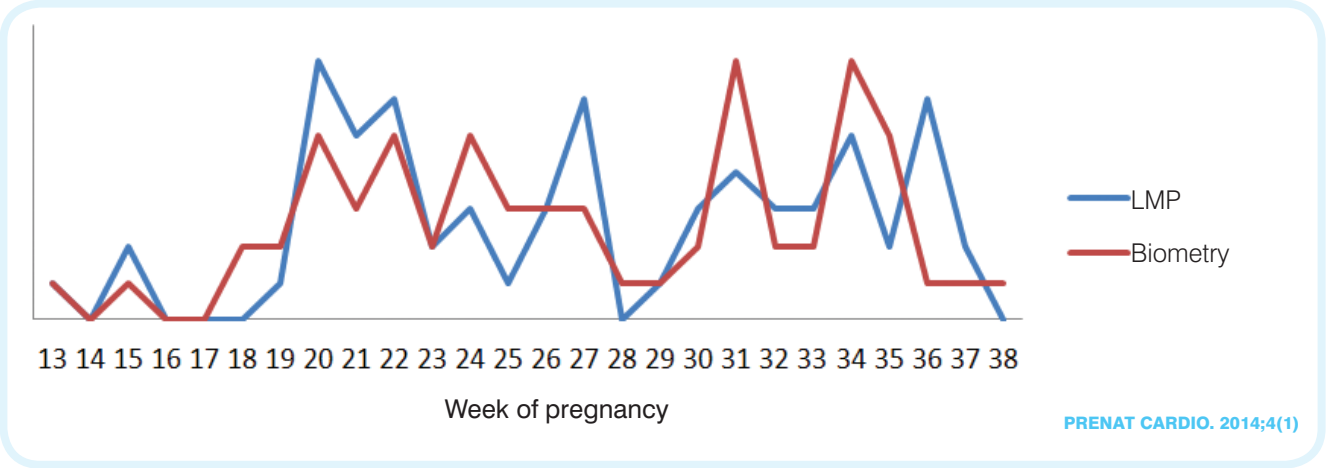

Figure 3. Nr of fetuses with PS since 13th week of gestation up to 38th week

pulmonary valve is usually tapered with poststenotic enlargement, may be present sinusoidal wave in the light of the oversized right ventricle. Proceedings of the newborn over the past decade has changed in most centers. In the center of Lodz the disadvantage qualify as a disadvantage urgent (critical), and we plan to balloon angioplasty in the first hours after birth. In the center of Warsaw attempts are made to art balloon in the fetus. A side from administration of prostaglandin in order to maintain the patency of the ductus arteriosus, balloon angioplasty may be considered in the newborn as isolated procedure, angioplasty with deferred treatment, the most common assumption palliative anastomosis BT, less the newborn can also consider correcting the total. Currently there are several publications on fetal pulmonary valve stenosis mainly relating to the diagnosis and prognosis, and the goal of our current analysis was to assess the quantitative prevalence of heart defect pulmonary valve stenosis in the Polish population of fetuses of pregnant women. In our database isolated pulmonary valve stenosis was on the 18th place of all heart defects, and database Allan and Sharland 20th place, which seems to suggest a similar prevalence: the Polish database of 1.4%, the UK 1.1% . On the other hand, our figures indicate that prenatal cardiac diagnosis in Poland is currently at a very good level, difficult to detect heart defects currently are often diagnosed by us more often than in the population of the UK in the late twentieth century.

CONCLUSIONS:

The incidence of isolated pulmonary valve stenosis in ORPKP database beetween year 2005 and 2013 rised by 1.3% and this value was similar to rates found in British Allan Sharland database. Isolated pulmonary valve stenosis on average was diagnosed relatively late, after 27 weeks of pregnancy, most often in nulliparous in low-risk pregnancies as a result of natural conception. Pregnancy after in vitro accounted for less than 10% of the total analyzed population of fetuses with isolated pulmonary valve stenosis.

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