

## SELECTED DATA FROM THE POLISH NATIONAL PRENATAL CARDIAC PATHOLOGY REGISTRY FROM THE YEAR 2016

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**Abstract**

**Introduction:** Analysis have been subjected to evaluate standard data reports from the Polish National Prenatal Cardiac Pathology Registry from the year 2016, compared to previous years.

**Material and methods:** The methodology of this work was comparison of the data from previous years that have been published in the Prenatal Cardiology and records generated in [www.orpkp.pl](http://www.orpkp.pl) from the year 2016. The total number of records entered in the Registry in the year 2016 was  $n=774$ .

**Results:** Top 10 most common prenatal CHD in 2016 were such as follow: HLHS, AVSD, VSD, d-TGA, TOF, AvS, muscular VSD, Aberrant origin R subclavian artery, RAA, CoA. In the centers Lodz and Krakow the most common prenatal congenital heart defects were severe CHD requiring surgery in 1 st month of life. In the contrast, in Warsaw the first place was taken by a "critical heart defects" regardless whether cardiac surgery was planned in first week (contemporary definition of prenatal critical heart defects) or first month - contemporary definition of severe planned heart defects of postnatal life.

**Conclusion:** The fact that in 2016 most common cause of referrals to targeted the fetal chocardiography was abnormal, large vessels view and not the abnormal 4 chamber view of the heart, suggests more and better training of physicians performing the screening or basic study of fetal heart in Poland.

**Key words:** Data base, Registry, Congenital malformation, Fetal echocardiography

**INTRODUCTION**

Analysis have been subjected to evaluate standard data reports from the Polish National Prenatal Cardiac Pathology Registry from the year 2016, compared to previous years<sup>1-5</sup>.

**MATERIAL AND METHODS**

The methodology of this work was comparison of the data from previous years that have been published in the Prenatal Cardiology and records generated in [www.orpkp.pl](http://www.orpkp.pl) from the year 2016. The total number of records entered in the Registry in the year 2016 was  $n=774$ . The number of records registered in 2004-2016 was  $n=7884$ . In 2016, the C-type centers (input in the Registry minimum 100 CHD per year) were centers from Warsaw  $n=287$  (37%), Lodz  $n=163$  (21%), Kraków  $n=138$  (17%) and Ruda Śląska  $n=121$  (14%). All C-type centers had 85% of all records in 2016.

A-type centers (input 10 or more prenatal CHD per year) in 2016 were: the resorts of Kraków/Wieliczka  $n=22$ , Katowice-1  $n=18$ , Katowice-2  $n=15$ , Szczecin  $n=14$  and Wrocław  $n=10$  (Map 1).

**RESULTS**

In 2016 the most common among fetal cardiac defects was HLHS ( $n=90$ ). It was like in previous years (2013  $n=70$ ; 2014  $n=92$ ; 2015  $n=71$ ). Over the years our Registry showed an increasing trend of prenatal detection and diagnosis of HLHS.

The second most common prenatal CHD was AVSD ( $n=57$ ), like in 2014 (AVSD  $n=48$ ), 2015 (AVSD  $n=66$ ), the third heart defect most commonly detected prenatally in 2016 was VSD ( $n=56$ ). In the year 2014 it was AvS ( $n=33$ ), and in the year 2015 the Tetralogy of Fallot ( $n=57$ ). (Table 1)

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LPLp.	2014 Type of CHD	Nr of cases		2015 Type of CHD	Nr of cases		2016 Type of CHD	Nr of cases	
1.	Hypoplastic left heart syndrome	92	12,30%	Hypoplastic left heart syndrome	72	8,60%	Hypoplastic left heart syndrome	90	10.4%
2.	AVSD: atrial & ventricular components (complete)	48	12.3%	AVSD: atrial & ventricular components (complete)	66	7.9%	AVSD: atrial & ventricular components (complete)	57	6.6%
3.	Aortic valvar stenosis	33	4.4%	Tetralogy of Fallot	57	6.8%	VSD	56	6.5%
4.	VSD	33	4.4%	d-TGA	39	4.7%	d-TGA	55	6.4%
5.	Aortic coarctation	32	4.3%	VSD	32	3.8%	Tetralogy of Fallot	52	6,00%
6.	Complete transposition of great arteries - d-TGA	32	4.3%	Aortic valvar stenosis	28	3.4%	Aortic valvar stenosis	41	4.7%
7.	Tetralogy of Fallot	26	3.5%	Aortic coarctation	25	3,00%	Muscular VSD	38	4.4%
8.	Left SVC persisting to coronary sinus	26	3.5%	Discordant VA connections (TGA) in complex heart defects	24	2.9%	Aberrant origin R subclavian artery	38	4.4%
9.	Aberrant origin R subclavian artery	23	3.1%	Left SVC persisting to coronary sinus	22	2.6%	Right aortic arch	31	3.6%
10.	Muscular VSD	23	3.1%	Muscular VSD	21	2.5%	Aortic coarctation	29	3.3%

Table 1. Top 10 most common prenatal CHD in Poland in 2014, 2015, 2016, based on the data from www.orkp.p

From 2012, we divided prenatal heart defects into the following groups: non-urgent, severe, critical and most severe, according to the anticipated clinical condition of the neonate and time of planned cardiac surgery<sup>6-9</sup>. In 2016 the most common diagnoses were severe heart defects -

46,8%, subsequent planned for cardiac surgery during 1<sup>st</sup> month after birth. Non urgent heart defects for surgery in infancy were 33%. For urgent invasive cardiac procedure as soon as possible - critical heart defects - 20% and heart defects – most severe- 0.2% (lethal cases). In the

centers from Karkow and Lodz in 2016, the most commonly diagnosed were severe heart defects (69%, 38,02%), differently the heart defects were classified in Warsaw, where most of CHD were critical heart defects (2016 – 41,98%, 2015-31%, 2014-30.5%), meaning old fashioned classification such as “ductal dependent”. So critical defect was meant that all CHD ductal dependent were considered the same, regardless would newborns required prostin or prostin and invasive procedure in cath lab. Otherwise there was no differentiation such as newborn baby needs treatment immediately after childbirth or later after 2-3 weeks (Table 2).

Fetal haemodynamic progression was observed in 2016 in 45 cases, in 2015 in 47 fetuses and 2014 in 62 fetuses.

In the year 2015 most common karyotype abnormalities were Down



Chart 1. Number of HLHS in Poland in www.orkp.pl in years 2004-2016

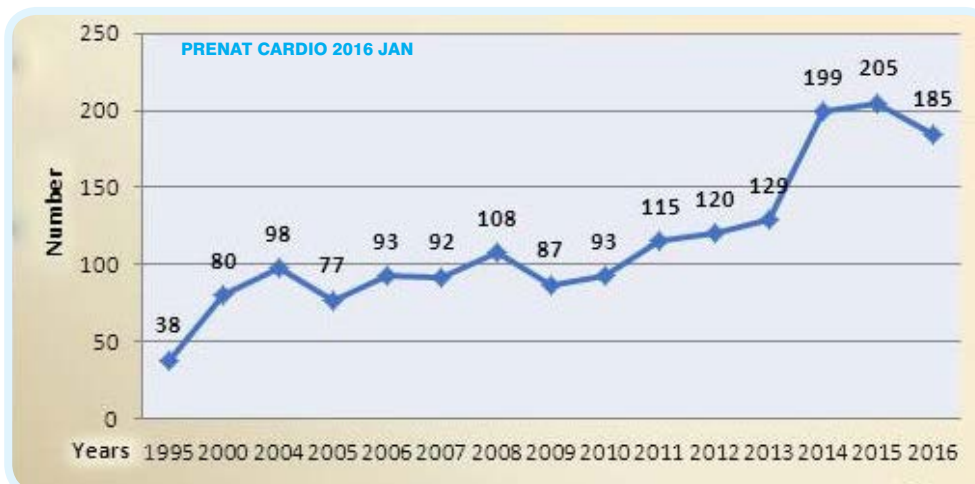


Figure 2. Publications from the scope of prenatal cardiology based on PubMed

Center	Nr of exams		Classification of CHD	Number	%
LSF	311	39.2%	CHD critical	106	30.55%
			CHD severe	92	26.51%
			CHD non - urgent	83	23.92%
			CHD most severe	9	2.59%
PBU	99	12.5%	CHD severe	161	69.4%
			CHD non - urgent	41	17.67%
			CHD critical	13	5.6%
			CHD most severe	2	0.86%
YBX	93	11.7%	CHD severe	46	38.02%
			CHD non - urgent	43	35.54%
			CHD critical	9	7.44%
			CHD most severe	0	
Center	Number of exams		Classification of CHD	Number	%
LSF	255	32.7%	CHD critical	94	31.44%
			CHD severe	86	28.76%
			CHD non - urgent	73	24.41%
			CHD most severe	1	0.33%
PBU	120	15.4%	CHD severe	236	76.13%
			CHD non - urgent	38	12.26%
			CHD critical	7	2.26%
			CHD most severe	4	1.29%
YBX	115	14.7%	CHD severe	55	44.35%
			CHD non - urgent	48	38.71%
			CHD critical	7	5.65%
			CHD most severe	3	2.42%
Center	Nr of exams		Classification of CHD	Number	%
LSF	263	32.1%	CHD critical	123	41.98%
			CHD non-urgent	96	32.76%
			CHD severe	61	20.82%
			CHD most severe	1	0.34%
PBU	148	18%	CHD severe	181	68.56%
			CHD non-urgent	47	17.8%
			CHD critical	16	6.06%
			CHD most severe	1	0.38%
YBX	105	12.8%	CHD severe	51	40.16%
			CHD non-urgent	40	31.5%
			CHD critical	17	13.39%

Table 2. Number of examinations of fetuses with CHD in different fetal tertiary cardiac centers in 2014, 2015, 2016

Abnormal karyotype	Number of cases	
Trisomy 18 - Edward's syndrome	19	37.3%
Trisomy 21 - Down's syndrome	17	33.3%
Other chromosomal anomaly	6	11.8%
22q11 microdeletion - CATCH 22	2	3.9%
Trisomy 13 - Patau's syndrome	2	3.9%
XXY - Klinefelter's syndrome	1	2%
Cat-Eye syndrome - Trisomy 22pter-q11	1	2%
45X0 - Turner's syndrome	1	2%

Table 3. The most common chromosomal abnormalities

syndrome n= 24 (50%) cases, the second place was Trisomy 18 n= (12,5%), the third was Trisomy 13 n= 6 (12,5%). In 2016, we notice a change in most reports of irregularities in the form of a karyotype Trisomy 18-Edwards syndrome n = 19 (37,3%), the second place was taken by the Down syndrome n = 17 (33.3%) cases. (Table 3)

In both 2014 and 2015, the most common cause of referral fetuses for fetal Echocardiography was abnormal 4 chamber view (2014 – 61.8%, 2015-61,2%) which is the basic step of fetal heart exam<sup>10</sup>. In 2016, the most common cause of referral for fetal echocardiography was the detection of abnormal great vessels view-43.2%, the second place was the detection of abnormal 4 chamber view – 33.6%.

In 2016 in majority of cases with congenital heart defects fetuses presented appropriate gestational age AGA – 88,1%. Small for gestational age (SGA) was present in 10,2% and large gestational age (LGA) was in 1,7%. In 2015 it was similar AGA – 89,6%, SGA-7,5%, LGA- 2,9%.

Amniotic fluid index in 2016 was normal in 93, (5%) of cases, polyhydramnion were 3,4% of the cases, and oligohydramnion was detected in 3,1%.

In 2016 decision about continuation of pregnancies despite of CHD was taken by 83,5% pregnant women. In 2015 such a choice was taken by 82,1%, and in 2014 by 85% women.

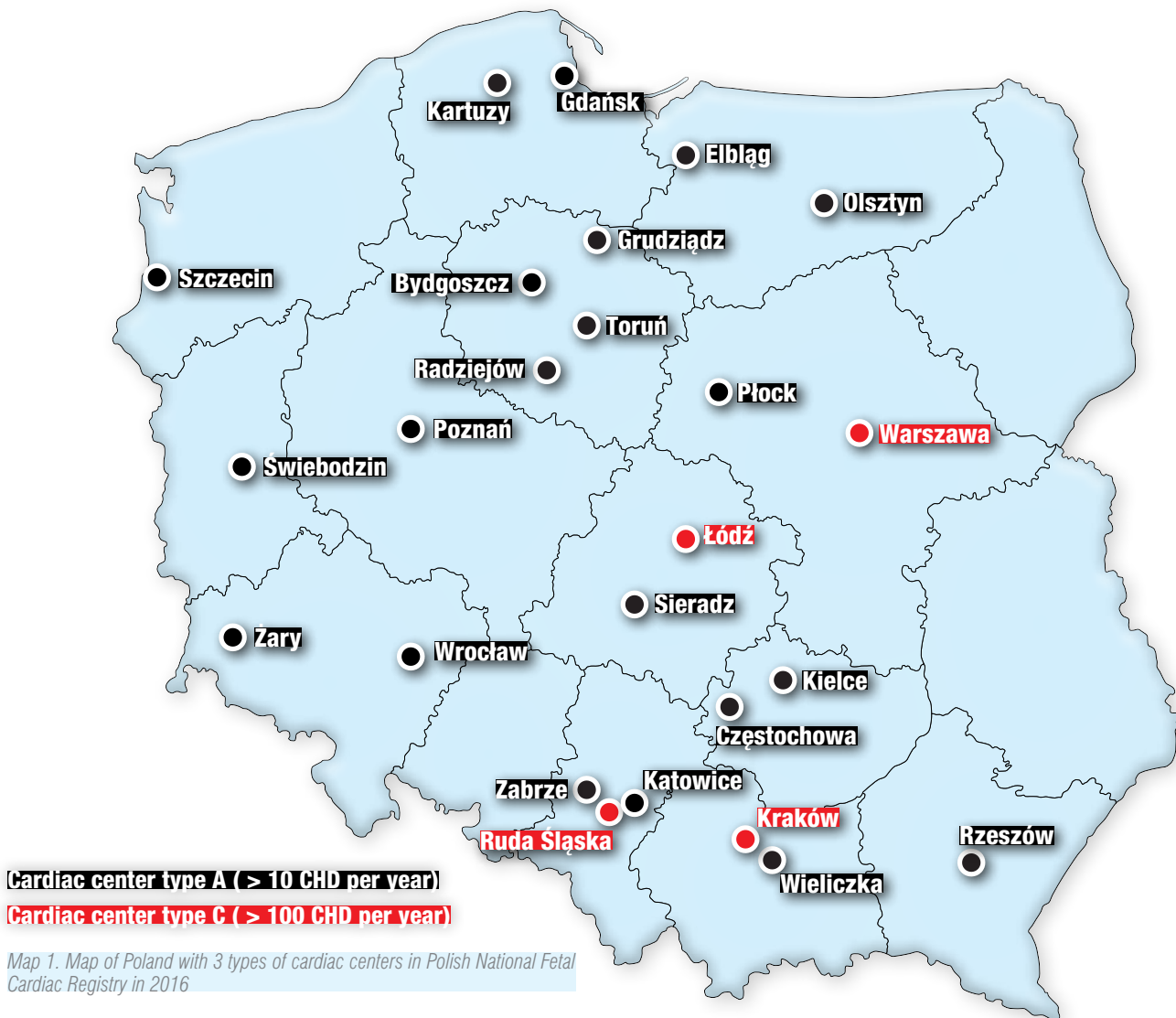
In 2016 there were 2 cases of intrauterine demises and in 2015 - 3 cases and in 2014 n=13.

In 2016, there has been a significant change in the labors way in favor of births with cesarean section: 70.4%, vaginal deliveries-29.6%. In 2015, CS-VD, 50%-50%, 2014 vaginal deliveries 56.1% of cases.

In 2016, the most frequently studied were fetuses between the 19-24 week of pregnancy i. e. . in 53,13%.

From 2014, we observe an increase in the number of medical centers cooperating with Registry: 2014 -25, 2015 – 29, 2016-28.

It is also noted that only small percentage of records were completed the forms for childbirth. They were filled in n = 85 cases in 2016, and a form for the diagnosis and treatment of postnatal n = 34. The number of records in ORPKP. pl, have



documentation of the detected CHD in the form of drawings, or frozen images or, "cine-loops". Input into the Registry is one of the main principles of applying for the Certificate of Professional Fetal Heart Examination in Poland<sup>11-13</sup>.

## DISCUSSION

By comparing the data from the year 2016 with data from previous years, we have noticed some similarities and differences. The largest centers of prenatal Cardiology remains: Warsaw, Lodz and Krakow: three C-type centers, introducing a min. 100 CHD to the Registry ([www.orpkp.pl](http://www.orpkp.pl)) per year.

The most commonly detected heart defect in 2016 remains HLHS (Hypoplastic Left Heart Syndrome)-like in previous years. The second place was occupied from 2014, in the form of a heart defect AVSD. In Warsaw the classification of heart defects of the fetus was different than in Krakow and Lodz, where the majority of the defects of the fetal heart, were critical and in other centers were severe.

In 2016, most patients were referred to the Echocardiography centers because of abnormalities in the image of large vessels, unlike in previous years, where abnormal 4 chamber view of the heart was the leading cause.

The most commonly detected abnormal karyotype of fetuses both in the year 2014, 2015 was Down syndrome, and in 2016, the first was Trisomy 18 Edwards syndrome  $n = 19$ .

In 2016, we can observe also a change the way of labor in favor of births with cesarean sections in relation to vaginal deliveries (70.4% to 29.6%).

Data analysis in subsequent years shows the direction of the development of prenatal cardiology in Poland. Also the number and the subject of publication in a digital database-PubMed, suggests the steady development of prenatal cardiology and different types of registries (Fig.2) at the global level, European and Polish level as well<sup>14-28</sup>. Although our National Registry is not perfect one and would need some improvement, still it is a unique organizational "tool" as well as prenatal cardiac defects collection.

We also would like to point the necessity of fetal heart examination in third trimester of the pregnancy<sup>29,30</sup>. This study is also very important and could have a strong influences on detection and prenatal diagnosis of CHD.

## CONCLUSION:

The fact that in 2016 most common cause of referrals to targeted the fetal chocardiography was abnormal, large vessels view and not the abnormal 4 chamber view of the heart, suggests more and better training of



physicians performing the screening or basic study of fetal heart in Poland.

Deliveries in pregnancies with fetal heart defects mainly by caesarean section (70% in 2016) is worrying, suggesting the need for a specialized prenatal heart program in the perinatal centers to help the obstetricians in making optimal decisions about how and when to give birth, because most heart defects of the fetuses and the newborns were clinically silent and did not require urgent treatment.

In Poland the development of prenatal cardiology is also proved by increasing of the number of prenatal cardiology centres of type A.

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#### Division of work:

**P. Kordjalik - first draft of the manuscript, analyzes of the data**

**Z. Tobota - work with the manuscript**

**M. Respondek-Liberska - final version**

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