● Original **paper**

HLHS ON THE BASIS OF A NATIONWIDE REGISTRY OF FETAL CARDIAC PATHOLOGY WWW.ORPKP.PL

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PRENAT CARDIO. 2013 SEP;3(3):18-21
DOI 10.12847/09133

Abstract

Congenital anomalies of development are an important issue in terms of both medical and social problems. In Poland in the years 2004-2012, an increase of detection of congenital heart defects in the form of HLHS (in 2004 - 22 fetuses, in 2006 - 38 fetuses, in 2008 - 66 fetuses, in 2010 - 69, in 2012 - 79 fetuses was observed). In 2012, the Nationwide Registry of Fetal Cardiac Pathology in the form of heart defect HLHS occupied first place among the most frequently detected heart defects, 10.2% of all registered patients with heart defects.

Key words: *congenital heart defects, hypoplastic left heart syndrome, registry of anomalies, Nationwide Registry of Fetal Cardiac Pathology (ORPKP), Nationwide Registry of Children's Congenital Anomalies (ORWWD), Eurocat, National Registry of Cardiacsurgeries Procedures*

In Poland, the most commonly detected developmental defects are congenital heart defects accounting for about 0.8-1% of live-born infants, and their pathogenesis is unknown in the majority of cases (90%).

Polish National Registry of Fetal Cardiac Pathology www.orpkp.pl, was created in 2004 as a pioneering concept for monitoring congenital heart disease in Poland. During the nine-year running period, the database has contributed to the assessment of the most frequently detected fetal cardiac pathologies.

The total number of records made in the years 2004-2012 was n = 4617, including the 1291 cases that were introduced into the registry by the Lodz center (PBU).

One of the most frequently detected heart defects is HLHS - hypoplastic left heart (Hypoplastic Left Heart Syndrome). This defect may concern all or part of the left heart structure: left atrium, mitral valve, left ventricle, aortic valve, ascending aorta, aortic arch, descending aorta.

HLHS can preexist with the premature closure of the foramen ovale valve, it can also be an isolated malformation or can occur as part of a genetic defect in the fetus (eg. trisomy 13).

How to cite this article:
Kordjalik P, Respondek-Liberska M. "HLHS on the basis of a Nationwide Registry of Fetal Cardiac Pathology www.orpkp.pl". Prenat Cardio. 2013 Sep;3(3):18-21

According to prof. Respondek-Liberska HLHS is one of the "simpler" to detect pathologies during screening, because with this defect, there is always an incorrect four camber view. As is clear from the reports listed in orpkp.pl, an abnormal four chamber view is one of the most common reasons for targeted fetal echocardiac examination and cause for referral to the type C center in Lodz (Department of Diagnosis and Prevention of Congenital Malformations ICZMP).*

The average number of examinations needed to detect HLHS in all the centers was 3, for the center of Lodz-2. The average gestational age at the time of detection based on biometrics and date of last menstrual period is 25 Hbd. It is known that the second trimester fetal ultrasound examination, between 18 - 22 weeks of pregnancy, is the most important in terms of prenatal cardiology, as the fetal heart is mature enough anatomically and functionally, allowing the evaluation of its structures and parameters. 25 weeks of gestation for cardiac diagnosis is relatively early in relation to term, ... but relatively late considering the possible option to exercise the right to termination of pregnancy.

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Submitted: 2013-07-09; accepted: 2013-08-30

Lp.	Congenital Defects	Total cases
1	*All anomalies*	100667
2	*Congenital Hearts Defect*	31488
3	HLHS	1072

Table 1. Number of congenital heart defects in Europe in years 2007-2011 (based on data from www.eurocat.com)

L.p.	European country	2007	2008	2009	2010	2011	Total
1	Austria	4	2	7	-	-	13
2	Belgium	6	6	10	7	11	40
3	Croatia	2	2	6	5	-	15
4	Denmark	1	0	1	3	2	7
5	France	16	11	16	12	17	72
6	Germany	3	9	5	2	7	26
7	Hungary	25	33	20	23	-	101
8	Irland	15	12	15	11	6	59
9	Italy	18	12	18	15	13	75
10	Holland	6	6	5	12	3	32
11	Norway	26	26	19	18	17	106
12	Spain	13	19	4	10	-	46

Table 2. Number of HLHS in selected european countries (based on data from www.eurocat.com)

L.p.	Diagnoses of heart defects	Number of cases			
		All centers in Poland		Lodz center (PBU)	
1	*Hypoplastic Left Heart Syndrome*	483	10,2%	153	11.5%
2	*Atrial & ventricular septal defect*	408	8,6%	115	8.6%
3	*Ventricular septal defect*	298	6,3%	76	5.7%
4	*Tetralogy of Fallot*	244	5,1%	71	5.3%
5	*Complete transposition of great arteries*	173	3,6%	63	4.7%
6	*Double outlet RV*	139	2,9%	55	4.1%
7	CoA – koarktacja aorty / *Aortic coarctation*	96	2%	44	3.3%
8	*Aortic valve stenosis*	147	3,1%	42	3.2%
9	*Common arterial trunk*	99	2,1%	41	3.1%

Table 3. Diagnoses of most common CHD in ORPKP in years 2004-2012 based on the data from www.orpkp.pl

According to data posted in the www.orpkp.pl, between 2004-2012, the number of points in the Apgar scale obtained by the largest group of live-born infants with hypoplastic left heart syndrome ranged between 8-10 points (in the first minute of life). The mean birth weight - at that time - was 2760g, with a maximum weight of 4500g, 50% of infants were transferred from the neonatal unit for cardiac center.

Analyzing the data presented (Figure 1) highlights the growing trend for the frequency of prenatal detection of congenital heart disease in the form of HLHS both in the center of Lodz (PBU) and throughout Poland.

Hypoplastic left heart syndrome, back in the 70's of the twentieth century was considered a lethal heart defect. Currently, 90% of cases are classified as severe heart defects, in which specialistic and multistage treatment is required. According to Polish legislation, the detection of this defect allows for the termination of pregnancy, up until the time when the fetus reaches viability outside the womb. Despite this drastic possibility, many parents decide to continue with the pregnancy. Early prenatal diagnosis, allows time to prepare-also in terms of psychological factors- for a difficult and time-consuming struggle for the life of their child. Another option rarely chosen by parents' is the decision to discontinue persistent therapy and transfer their child into home hospice care.

Infants with HLHS in Poland and in Europe cannot be candidates for heart transplantation due to the shortage of donor organs, but it is possible in the U.S. and Canada. Based on real events concerning two married couples (one expecting a baby with HLHS and the other a newborn with a central nervous system congenital malformation) the U.S. film titled "Heart of the Child" was produced, it describes such dramatic events but of course with a happy ending.

We caution not only readers of Polish Prenatal Cardiology, but also the Ministry of Health, Ministry of Finance, managing on all levels of society, that financing the diagnosis and treatment of our patients should take into account the growing trend of deliveries of newborns with heart defects, especially of the defect in the form of HLHS.

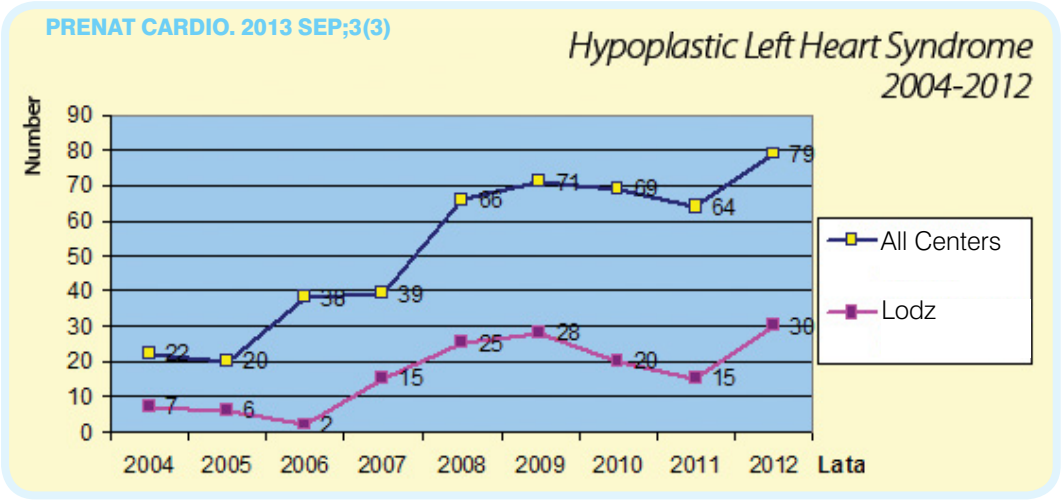

Chart 1. Graphical presentation of number of fetuses with HLHS in years 2004-2014 (based on www.orpkp.pl) in Poland and in one center Lodz (PBU)

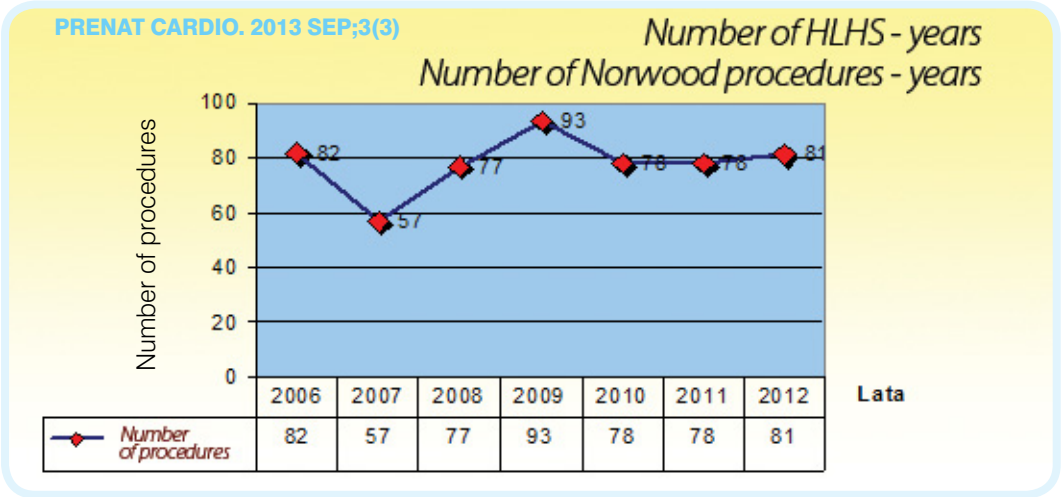

Chart 2. Graphical presentation of number of Norwood procedures in neonates in Poland in years 2006-2012 (based on data from www.krok.org.pl)

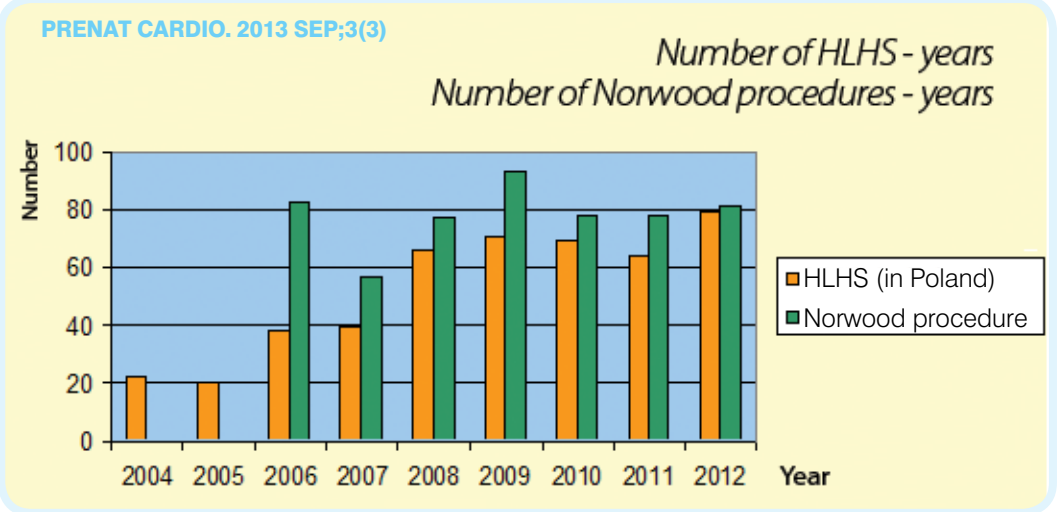

Chart 3. Graphical presentation of number of fetuses with HLHS and number of neonatal Norwood procedures in Poland (based on www.orpkp.pl and www.krok.org.pl)

L.p.	Neonatal data	All centers	Lodz center (PBU)
1	Number of fetuses with HLHS	437	143
2	Mean number of US until anomaly was detected	3	2
3	Mean gestational age at the time of anomaly detection (in wks) base on last menstrual period	25	25
4	Mean fetal gestational age at the time of anomaly detection based on fetal biometry	25	25
5	Apgar score 9-10 in the forst min of postnatal life	46,4%	56,3%
6	Mean birth weight (g)	2760	3017

Table 4. Basic parameters of fetuses and neonates with HLHS in years 2004-2012 (based on data from www.orpkp.pl) .

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Financing: The research was not financed from the external sources
Conflict of interest: The authors declare no conflict of interest and did not receive any remuneration