DOES EVOLVING FETAL HEART DEFECTS POSE A SEPARATE CLINICAL PROBLEM ?

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Abstract

A retrospective analysis of medical records of selected 83 cases (pregnant women and fetuses of patients from the database of the Department of Diagnosis and Prevention of Congenital Malformations ICZMP in Lodz in the years 2007-2012) who had minimum 2 echocardiographic exams. In this group 220 echocardiography exams were performed: in 62 fetuses 2x and in 21 fetuses 3x or more. After exclusion of intrauterine demises and terminated pregnancies, 72 cases have been qualified for further analysis. Heart defects in this material were initially divided into four groups: the most serious defects, critical, serious and evolving. Group of the most serious defects was excluded from the 2nd stage of analysis. An attempt to subtract the group of "evolving" heart defects from the group of severe and critical defects was conducted. This group of defects was initially classified as "severe evolving to the critical" , as hemodynamics progression was observed during intrauterine life . Mortality in the subgroup of evolving defects was 71.4 %, in the group of critical defects 36.8 % and in the group of heavy defects 34.4 % (chi-square test 0.05). In the group of evolving defects the mortality rate after cardiac surgery was :7/11 infants (63 %) and in the group of severe defects 7/26 (26.9 %) (Fisher test 0.018). There were no other differentiating features found within the statistical analysis of the study groups.

Conclusions: Prenatal classification of fetal heart defects based on echocardiographic monitoring allows to observe the progression of hemodynamic changes in selected fetuses. This group of defects was encumbered with the greatest mortality in the neonatal and postoperative period.

Key words: *classification of congenital heart defects, severe defects, critical defects, progression of hemodynamic changes*

INTRODUCTION

In the past few decades we have witnessed a rapid development of prenatal diagnoses [1,2]. In Poland the first prenatal ultrasound was performed in 1975. Congenital heart defects are the most common birth defects and occur in 0.8 - 1% of live-born infants [3,4,5,6,7] . Fetal heart defects are three times more likely, however, as many as 60 % die before birth [8,9]. Most of the defects are asymptomatic, both during pregnancy and immediately after birth and even during the first days of life [10,11,12].

Currently, according to the National Registry of Cardiac Problems in Fetuses, the five most common defects are the following: hypoplastic left heart syndrome (HLHS) - 11.3%, atrioventricular septal defect (AVSD) - 7.6% ; tetralogy of Fallot (TOF) - 6.1 %; ventricular septal defect (VSD) - 5.1% ; transposition of great arteries (d - TGA) - 4.3%.

Those defects account for approximately 34 % of all fetal heart defects (based on national data on more than 4,000 fetuses in the years 2010-2013) [13].

The development of prenatal echocardiography launched the new divisions of heart defects, initially from three- into four groups [14,15](Table 1).

This paper attempts to extract from severe and critical congenital heart defects a subgroup, which was titled "evolving defects". The group of evolving malformations was distinguished on the basis of changes in hemodynamics during intrauterine life.

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Type of heart defect	*Procedures*
„Mild" defects	*Non-life-threatening condition for fetus or neonate, usually planned surgery in infancy or later.*
Severe defects	*Threat to life of the newborn, usually ductal dependent circulation (after delivery prostin required to keep ductus arteriosus opened); elective neonatal cardiac surgery*
Critical defects	*Urgent procedure required just immediately after delivery or during the pregnancy (balloon valvuloplasty and/ or balloon atrioseptostomy and subsequent cardiac treatment)*
The most severe defects	*No chance for survival (for fetus or neonate) in the current state of development of medicine[14,16].*

Table 1. Basic four types of heart defects in prenatal cardiology (classification for obstetricians and neonatologists)

"Evolving Fetal Heart Defects"		
Hemodynamic changes		***Number of fetuses***
Restrictive FO		8
Progression of abnormal (reversal) flows through the pulmonary veins		4
Functional tricuspid regurgitation		2
Progress in oligohydramnios		2
Intrauterine growth restriction		2
Increasing fetal heart size (cardiomegaly)		1
Others:	*Pericardial effusion*	1
	Hyperechogenic bowels	2
	Generalized edema of the fetus	1
	Abnormal venous flows	2
	Thickened placenta (> 6 cm)	6

Table 2. New subgroup of fetal cardiac defects: "Evolving Fetal Heart Defects" (n = 14)

AIM OF THE STUDY

Analysis of data regarding evolving fetal heart defects based on echocardiographic monitoring in the reference prenatal cardiology center.

Is the fate of fetuses with evolving heart defects different from that of fetuses with severe and critical heart defects?

MATERIAL

The study group - pregnant women who underwent at least two fetal echocardiographic examinations in the Department of Diagnosis and Prevention of Congenital Defects ICZMP in 2007-2012 , on the basis of which the prenatally diagnosed congenital heart malformation was initially classified as a severe or critical heart defect, occurring in singleton pregnancy, with complete medical records accumulated in a computer database (FMaker Pro, Microsoft), on fetal cardiac examination and course of pregnancy and on the newborn.

METHOD

A retrospective analysis of medical records of selected 83 pregnant women who underwent 220 echocardiograms of the fetal heart, in that in which 62 fetuses were examined twice, 21 fetuses of 3 or more times. Disqualified from the analysis were fetuses with heart defects who underwent echocardiography only once, multifetal pregnancies, fetuses with chromosomal aberrations and fetuses that underwent invasive therapy in utero.

Methods of statistical analysis. For the analysis of the collected data, the following statistical methods were used :

chi-square test of independence or Fisher's exact test [17];

way analysis of variance with multiple comparison test for comparing means (for continuous variables), in two or more groups[18] .

An analysis of the follow up of 83 fetuses with heart defects was conducted . Termination of pregnancy was conducted in 6 cases (7.2%) , death " in utero " concerned 5 cases (6.1 %).

For further analysis a group of live-born infants was qualified, from which a subgroup of infants with the severest congenital defects were eliminated n = 7 (expected death, conservative proceedings) .

The group comprised of evolving defects did not differ in terms of structural abnormalities of the heart, from the other groups. It was a group, which contained the same anatomical defects, occurring in the group of severe and critical defects. Hemodynamic monitoring and classification of them as severe malformations with the possibility of deteriorating into a critical state gives evidence of the dynamism and evolution of changes that occur during fetal life and were recorded on echocardiography (Table 2) .

Table 3. A comparison of the type of pregnancy in three groups of heart defects: critical, severe and "evolving"

Type of pregnancy	Critical defects		Severe defects		Evolving defects	
	n	%	n	%	n	%
High risk pregnancy	5	26,3	11	34,4	1	7,1
Low risk pregnancy	14	73,7	21	65,6	13	92,9
Total	19	100	32	100	14	100
0,162 *P in the Fisher's exact test*						

Frequency distributions (High risk pregnancy and low risk pregnancy based on maternal medical history) within different types of heart defect groups were compared by Fisher's exact test (Fisher statistic = 3.714, p = 0.162).

Type of pregnancy	Critical defects n= 19 (100%)		Severe defects n= 32 (100%)		Evolving defects *n= 14 (100%)*	
	n	%	n	%	n	%
Vaginal delivery	6	31,6	12	37,5	1	7,1
Cesarean Section	13	68,4	20	62,5	13	92,9

Frequency distributions in the three types of heart defects were compared by Fisher's exact test (Fisher statistic = 4.536 , p = 0.106).

The remaining research compared statistics regarding structure of delivery in the group of evolving heart defects (type of pregnancy, form of delivery, week of delivery) with the structure of those in the group of severe and critical defects. Frequency distributions among different types of groups were compared by chi -square test of independence. In all analyzed groups of heart defects term deliveries were similar and amounted to over 60 % (Fisher statistic = 0.757 , p = 0.999). The Apgar score of more than 7 points in the analyzed groups of heart defects were similar and amounted to 79% of critical defects , severe -66 % and 86 % of defects evolving. These values did not differ significantly (Fisher statistic = 0.304 , p = 0.999) . If we take into account birth weight, significantly more infants with low birth weight < 1000 g occurred in the severe defects group (Fisher statistic = 7.393 , p = 0.063). Finally, compared to the incidence of death in all three groups of heart defects (Table 6). The scale of demise among evolving defects (71.4%) was twice as frequent as the mortality among critical (36.8 %) and severe defects (34.4%) (χ 2 = 5.879 , two st.sw. , p = 0.05). This group was characterized by the highest mortality rate following cardiac surgery (p = 0.018).

DISCUSSION

Severe and critical heart defects in fetuses are a difficult clinical problem , more so because in some cases the haemodynamic condition of the fetus may vary [19,20,21,22]. Observation and analysis of individual cases allowed the distinction of defects classified as severe " turning " into critical. These defects are called evolving. Amongst severe defects in the fetus, there is usually a certain amount of time available in order to take action after delivery: the administration of medication counteracting fetal states which may directly cause haemodynamic deterioration in the newborn - even deferred for one month. In contrast, critical defects of the fetus require proceedings within the first 24 hours of life or immediately after birth, or sometimes even in intrauterine life. In the case of evolving defects deterioration of the fetus or newborn may come as a surprise to the medical staff , because infants with a heart defect, whether critical or severe, both receive a similar Apgar score. The problem was evident by the percentage of neonatal deaths : the evolving defects

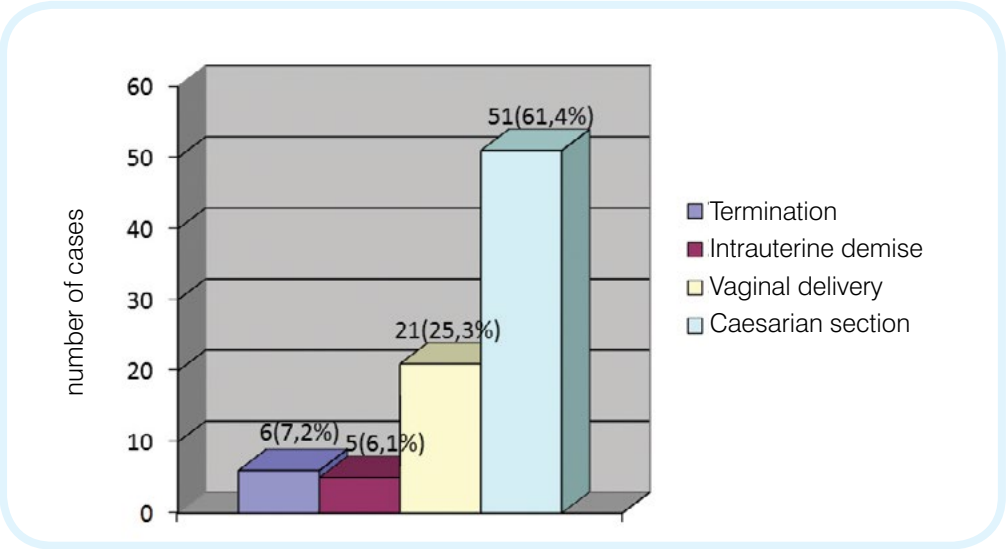

Figure 1. The total number of cases in the study n = 83 (100%) (Forces of nature= Vaginal delivery)

Table 5. Parameters of the newborns in the three groups of heart defects

Type of heart defect according to the criteria of prenatal diagnoses	***Date of birth***		***Apgar score***		***Birth weight***		
	< 37 wks	37 wks	< 7 pkt	7 pkt	≥2500g	< 2500g	< 1000g
Critical defects, n=19	6 (32%)	13 (68%)	4 (21%)	15 (79%)	17 -90%	2 -10%	0
Severe defects, n=32	11 -34%	21 -66%	11 -34%	21 -66%	12 -38%	17 -53%	3 -9%
„Evolving defects", n= 14	5 (36%)	9 (64%)	2 (14%)	12 (86%)	13 (93%)	1 (7%)	0
The probability of a comparison test frequency distributions		0,999		0,999			0,063
Total n= 65(100%)	22 (34%)	43 -66%	17 (26%)	48 (74%)	42 (64%)	20 (31%)	3 -5%

Frequency distributions within different heart defect groups were compared by Fisher's exact test .

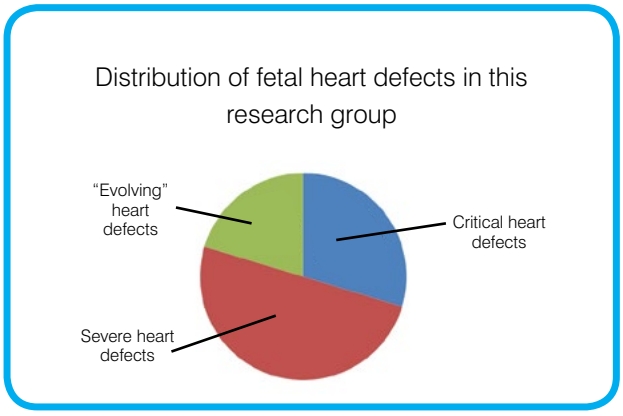

Figure 2. Distribution of fetal heart defects in this research group n = 65 cases (without the most severe defects – those with no chance for survival)

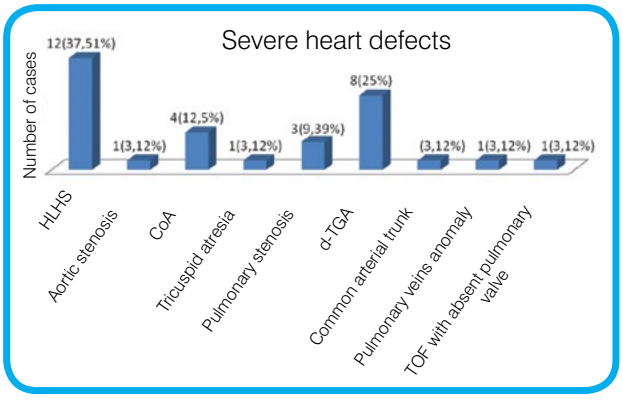

Figure 4. Distribution of severe heart defects (predicted possibility of survival after delivery based on elective cardiac surgery) in the analyzed population n = 32 cases (100 %)

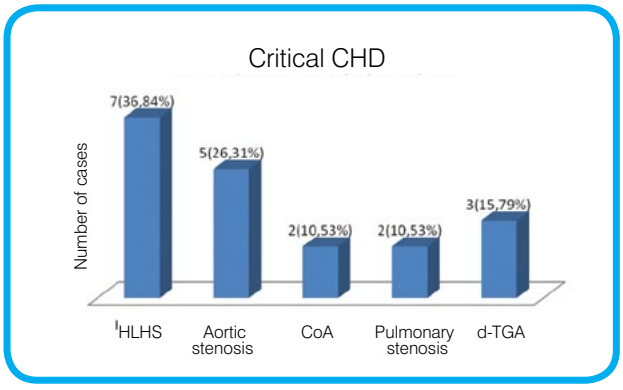

Figure 3. Distribution of critical heart defects (predicted possibility of survival after the application of therapy immediately after delivery or during pregnancy) in the analyzed population of n = 19 cases (100%)

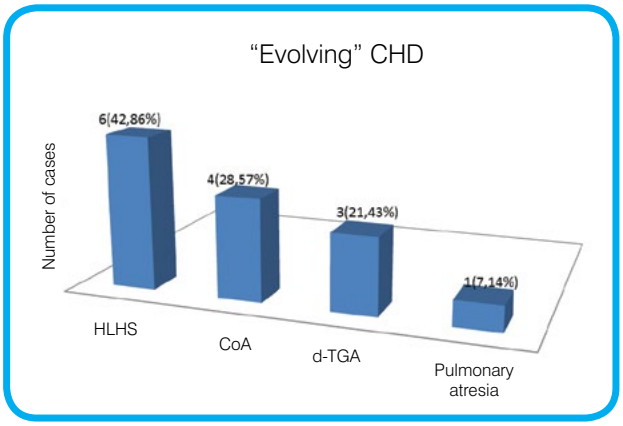

Figure 5. Distribution of "Evolving" heart defects (based on fetal echocardiography monitoring haemodynamic progression and deterioration) in the analyzed populations n=14 cases (100%)

was 71.4 %, with 36.8% of the critical defects and 34.4 % for severe defects (p = 0.05). A very important issue was the lack of any relation to birth weight, duration of gestation and Apgar score. Birth weight of infants with evolving defects in 93% was above 2500g compared to 90 % of neonates with critical defects. A similar case was presented in comparison to the duration of pregnancy. Labor-delivery time was over 37 weeks in the case of critical defects 68 %, evolving defects 62 % and 60% severe defects. The Apgar score adds even more " confusion " , as it was the group of newborns with evolving defects that constituted the largest group of ratings above 7 points . - Up to 93 % compared to 90 % of critical defects and 66% of severe malformations. On the basis of statistical analysis conducted amongst the study group of newborns, no features differentiating the division of fetal heart defects were found. The welfare of the newborn at birth , the existing criteria (Apgar score , birth weight) were not differentiating factors for the defects. However, the results of comparing the incidence of death in the perinatal period of severe defects , critical and evolving were statistically significant ($\chi 2 = 3.860$, df = 1, p = 0.05) and mortality after cardiac surgery (p = 0.018). It is this higher mortality amongst newborns in the evolving defects group that is remarkable.

Table 6. Comparison of the incidence of deaths in each group of heart defects

Comparison of the incidence	**Type of heart defect**					
	Critical		*Severe*		*„Evolving"*	
	n	%	n	%	n	%
Survival	12	63,20%	21	65,60%	4	28,60%
Death	7	36,80%	11	34,40%	10	71,40%
Total	19	100	32	100	14	100
p = 0,05 *P in a test comparing frequency distributions*						

Chi -square test of independence, at a significance level of 0.05, the value of Chi-square = 3.860 df = 1 , P = 0.05

CONCLUSIONS

The evolving defects group demonstrated the greatest mortality in both the perinatal and post-operative period.

One-time echocardiography in fetuses with congenital heart defects seems to be a rarity in prenatal cardiology.

Prenatal classification of fetal heart defects should include monitoring of fetal echocardiography in referral centers for prenatal cardiology.

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Table 7. Deaths after cardiac surgery in the neonatal or infancy period

Type of surgery	**Number of newborns**	**Death**		**Survival**	
		n	%	n	%
Norwood's operation	20	11	55	9	45
Crafford's operation	1	0	0	1	100
Jatene`s operation	10	2	20	8	80
Jatene`s in Lecompte's modification	1	0	0	1	100
Hemi-Fontan's operation	1	1	100	0	0
Bidirectional Glenn's operation	2	0	0	2	100
Pulmonary artery banding	2	1	50	1	50
Shunt implantation and atriospetostomy	2	1	50	1	50

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Table 8. Death and survival after interventions and without the interventions of cardiac surgery in three groups of heart defects

Follow-up procedure performed on the newborn		Critical defects		Severe defects		"Evolving defects"	
		n	%	n	%	n	%
Interventions procedures	Survival	12	66,7	19	73,1	4	36,4
	Death	6	33,3	7	26,9	7	63,6
Without the intervention	Survival	0	0	2	33,3	0	0
	Death	1	100	4	66,7	3	100
Total	Survival	12	63,2	21	65,6	4	28,6
	Death	7	36,8	11	34,4	10	71,4

Deaths and survival in each group after cardiac surgeries by Fisher exact test ($p=0,018$)

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