POSTNATAL OUTCOMES OF CHILDREN WITH PRENATALLY DIAGNOSED CONGENITAL HEART DISEASE COMBINED WITH CONGENITAL DIAPHRAGMATIC HERNIA

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Abstract

The primary aim of this study was to determine the predictive value of prenatal ultrasound and echocardiography for prognosis in congenital heart disease (CHD) with coexisting diaphragmatic hernia (DH) in a tertiary care center. Eleven records from the database of the Department for Diagnoses and Prevention of Congenital Malformations, Polish Mother's Memorial Hospital Research Institute, were reviewed. The mean maternal age was 29,2 ± 5,1 years, and the mean gestational age at the time of diagnosis was 28,4 ± 6,7 weeks. No information was available for children discharged from hospital. Data of eight cases of prenatal DH and complex heart disease from the literature were also analyzed. Three fetuses survived the neonatal period. In each of these, CHD was other than the urgent or critical type, defined as not requiring cardiac surgical intervention in the first day or month of life. Both sets of data collected from our center and the published literature confirmed the unfavorable prognosis for fetuses with severe or critical CHD with coexisting DH.

Key words: *Diaphragmatic hernia, complex congenital heart defect, prenatal diagnosis*

INTRODUCTION

Congenital diaphragmatic hernia is a severe pathology leading to pulmonary hypoplasia and pulmonary hypertension. Respiratory distress is considered as the major cause of death in neonates suffering from CDH.

Coexisting CDH and congenital heart disease increase the risk of serious complications and neonate's demise.

In the present report, we posed the question: is prenatal diagnosis using ultrasound adequate to predict the prognosis of congenital heart defects (CHD) with coexisting diaphragmatic hernia (DH)?

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MATERIALS AND METHODS

In the database from our unit (Filemaker Pro), all cases occurring between January 2007 and July 2014 and satisfying the following criteria were reviewed: diaphragmatic hernia and complex heart disease in singleton pregnancy and prenatal diagnosis, established at the Department of Diagnosis and Prevention of Congenital Malformations, Polish Mother's Memorial Hospital Research Institute (Table 1). The outcome of pregnancy was verified in both the Polish National Registry of Fetal Cardiac Pathology and Hospital's database.*

In the Medline database, publications concerning analogous case and case series reports were also analyzed (Table 2).

RESULTS

Data of the 11 cases from our unit are listed in Table 1. The mean maternal age was 28,0 ± 5,2 years, and the mean gestational age at the time of diagnosis was 29 ± 7 weeks.

The graphical representation of distribution of the gestational age of each fetus is shown in Figure 1.

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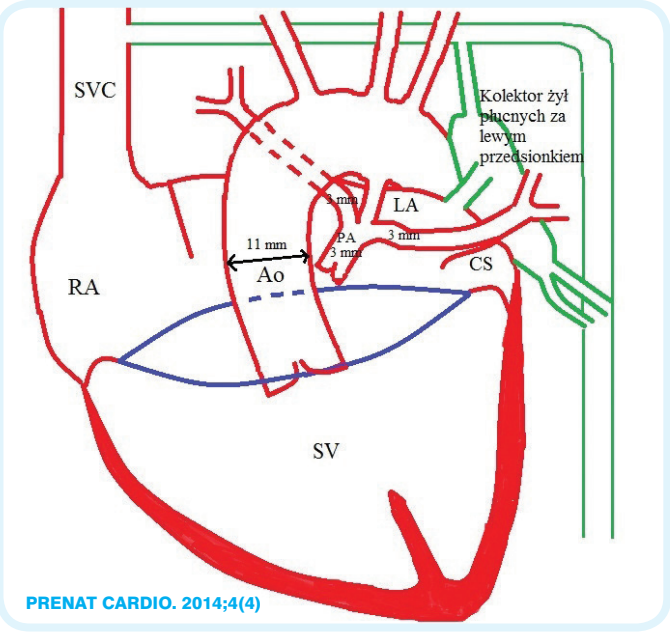
In two cases, pregnancy was terminated in accordance to Polish legislation. In five cases, neonates were delivered by caesarean section, and in one case by vaginal delivery, but with death on the first day of life. No information about the other three live-born neonates was found.

Eight cases from literature (Table 2) pertaining to prenatal diagnosis of complex heart defect with coexisting DH were analyzed. Termination of pregnancy was performed in three cases, four neonates died in the first days of life (in case Nº 6 parents decided to withdraw support of extracardiac oxygenation). The only newborn who did not required a cardiothoracic surgery in the first month of life remains alive and well. This child was not ductal dependent and had a planned later cardiac surgery.

DISCUSSION

The relation between DH and CHD has been well documented. Their coexistence is attributed to the formation of the diaphragm and heart at similar times during fetal development. CHD, among which hypoplastic left heart syndrome is the most frequent, are noted in 10 to 25 % of DH, especially in the left-sided DH[1].

Figure 1. Schematic drawing of the fetal heart defect with diaphragmatic hernia (case nr 11, table 1)



No	Year	CHD	Type of CHD in fetal cardiology	ECM	Fetal weight (in grams) at the time of diagnosis	Wks of gestation at the time of diagnoseses	Delivery	Geste age at the time of delivery	Neonatal birth weight (in grams)	Follow-up
1	2007	DORV, VSD	Non-urgent	DH, Galen Malformation, Polyhydramnion	2843	37	CS	39	2900	Death on 2nd day
2	2007	AV-canal + VSD musc., dextropositio	Non-urgent	DH, Single umbilical artery, IUGR, Ahydramnion	964		bd	bd	bd	No data
3	2008	AVC,	Non-urgent	DH, Lung hypoplasia, Hydronephrosis, Polyhydramnion, Megaureter	2920	38	CS	40	2980	Death after delivery (1h)
4	2009	Dextropositio, Disproportion, HLHS	Severe / planned	DH, polyhydramnion	881	27	CS	39	3150	Death on 6th day
5	2009	Dextropositio, AV-canal	Non-urgent	DH, hydrothorax, Polyhydramnion, SGA	1976	35	CS	35	2450	Death on 1st day
6	2009	TGA, HLHS/AV-canal unbalanced, Dextrocardia	Severe/ planned	DH, Polyhydramnion	1289	30	bd	bd	bd	No data
7	2012	Single ventricle, Trunus arteriosus, Dektrowersja, pericardial effusion	Severe/planned	DH, Diaphragma agenesis	316	19	CS	38	3800	Death on 1st day
8	2013	Tricuspid valve insuff., Hypoplastic aortic arch, disproportion	Severe/planned	DH – Right, polyhydramnion	1535	30	-	-	-	Death in utero
9	2014	Dekstrowersja, common AV-canal	Non-urgent	DH –Left	270	21	-	-	-	TOP (?)
10	2014	SV/Truncus , Pericardial effusion. Myocardial hypetrophy	Severe/planned	DH, Hydrothorax	300	22	-	-	-	TOP (?)
11	2014	AVC/SV + TGA + PA + TAPVC + CS	Severe/planned	DH	2216	38	SN	39	2410	Death on 2nd day

bd = no data

Table 1: Data from Department of Congenital Malformations Research Institute Polish Mother's Memorial Hospital regarding 11 fetuses with congenital heart defect and diaphragmatic hernia (years 2007-2014)

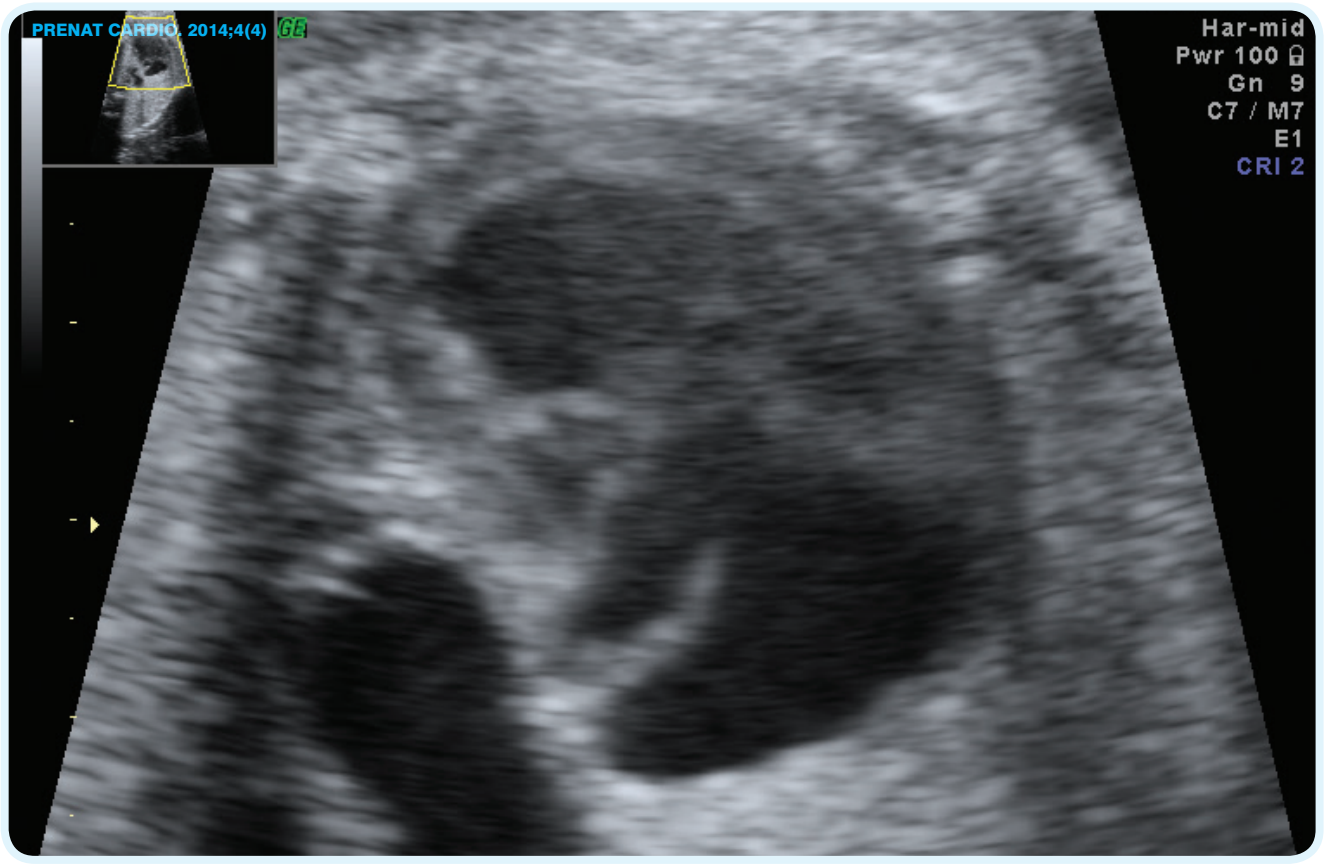


Figure 2. Congenital heart defect with dilated coronay sinus and stomach in the fetal chest (case nr 11, Table 1)

According to the analysis carried out in Polish Mother's Memorial Hospital Research Institute in 2006, diaphragmatic hernia coexisting with extracardiac malformations has been lethal. No patients diagnosed with DH and congenital heart disease survived beyond the neonatal period. [2]

As other authors reported, the prognosis in DH and CHD is poor[2-6]. No new scientific reports regarding DH+CHD have been observed since 2006. Also the late gestational age of the patients referred to our Department with respect to the consultations described in published reports should be emphasized, with regard to probable in utero demise of fetuses with these combined conditions .

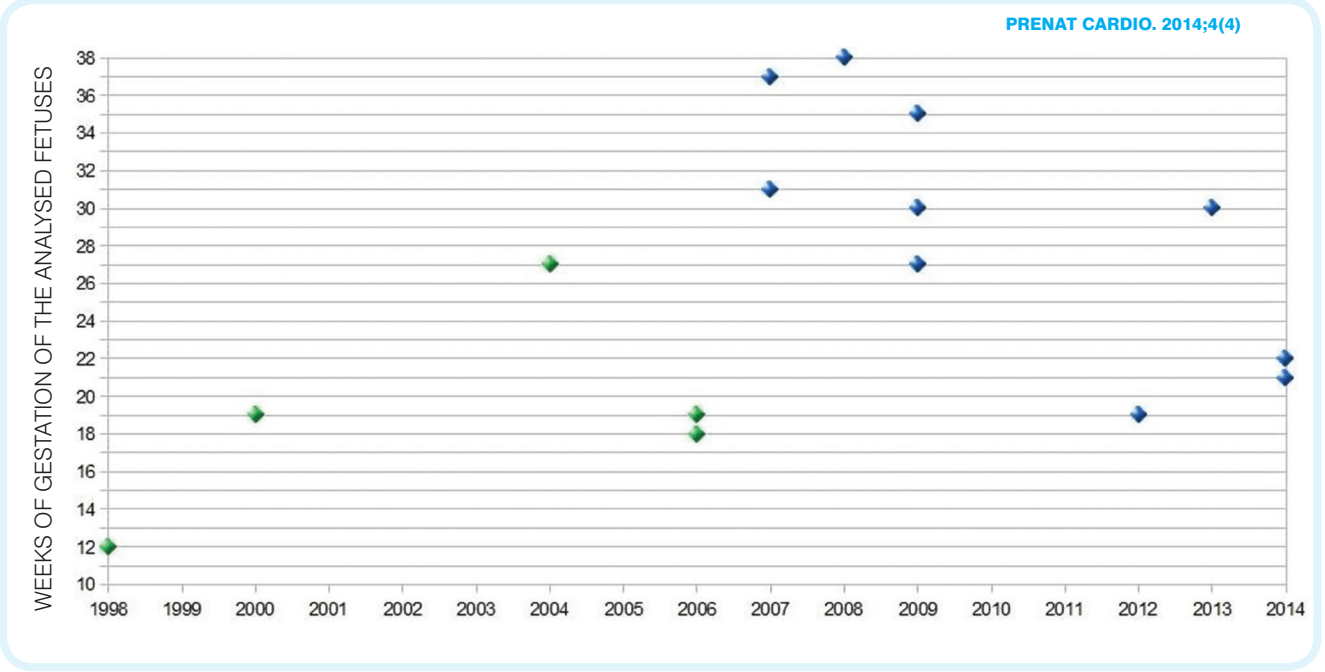


Chart 1. Presentation of the gestational age of fetuses with diaphragmatic hernia and congenital heart defect (from literature in green, from our uni in blue)

Lp.	Year	Author /Hospital	CHD + ECM	Type of CHD in prenatal cardiology	WKs of gestation at the time of diagnosis	Delivery	Birth weight	WKs of gestation at the delivery	Follow-up
1	1994	Respondek ML, Binotto CN, Smith S, Donnenfeld A, Weil SR, Huhta JC; Pennsylvania Hospital, Philadelphia, USA	VSD+DH+ dekstrokardia	Non-urgent	bd	-	-	-	Neonatal demise
2	1998	Yung Hang Lam, Mary Hoi Yin Tang, Siu Tsan Yuen, Hongkong, China	CHD, membranous VSD, DH (left side), hipertrofia RV, pulmonary valve atresia	Severe / planned	12	-	-	-	TOP
3	1999	Lucia Migliazza, Christian Otten, Huimin Xia, Jose I. Rodriguez, Juan A. Diez-Padro, Juan A. Tovar, Madrid, Spain	DH + TGA	Severe / planned	bd	-	-	-	Neonatal demise
4	2000	Lee Noimark[1], Mark Sellwood[2], John Wyatt[3] and Robert Yates[4] [1]University College London Medical School, London, UK; [2]Neonatal Intensive Care Unit, University College Hospital, London, UK; [3]Neonatal Intensive Care Unit, University College Hospital, London, UK; [4]Cardiothoracic Unit, Great Ormond Street Hospital, London, UK	TGA+VSD+DH, Diproportion	Severe / planned	19	-	-	37	8 month alive & well
5	2004	Hamrick SEG, Brook MM, Farmer DL, San Fransisco, California, USA	DH + pulmonary sequestration + TGA	Severe / planned	27	CS (EXIT)	2000 g	34	Demise
6	2006	Isabel López, Juan A. Bafalliu, M. Carmen Bernabé, Francisco García, Miguel Costa and Encarna Guillén-Navarro, Argentina	DH, AVSD	Non-urgent	18	-	-	-	TOP
7	2006	Isabel López, Juan A. Bafalliu, M. Carmen Bernabé, Francisco García, Miguel Costa and Encarna Guillén-Navarro, Argentina	IUGR, CHD+DH (left side), overlapped toes	-	19	-	-	-	TOP
8	2008	Wojciech Fendler, Andrzej Piotrowski, Medical University Hospital, Lodz, Poland	DH + d-TGA (in autopsy)	Severe / planned	bd	CS	bd	36/37	Death 1 h after delivery

bd = no data

Table 2: Data from literature: fetal heart defect + diaphragmatic hernia (1994-2008)

The Fetal Heart Team in Polish Mother's Memorial Hospital Research Institute is composed of specialists of different sectors of perinatal care to discuss diagnoses and treatment of the youngest patients – fetuses. During Fetal Heart Team meetings, we take into account the recent classification of congenital heart diseases proposed by our team of prenatal cardiologists[7]. In this classification there are:

Non-urgent heart defects (demanding intervention during infancy),

Urgent but planned ductal dependent (not immediate) (requiring classic cardiac surgery during the neonatal period)

Critical (usually also ductal dependent) heart defects for which immediate intervention after delivery or even *in utero* is required for;

The most severe heart defects meaning the treatment is currently not possible.

This primary classification of CHD was further changed by Słodki[8], who added two other categories of CHD: heart defects coexisting with structural extracardiac malformations (which may be potentially corrected by surgical intervention) and heart defects coincidental with extracardiac anomalies (they are not qualified to surgical treatment).

According to Słodki, only 14% of 89 neonates admitted to Polish Mother's Memorial Hospital Research Institute with CHD complicated by extracardiac anomaly survived the first month of life, provided that only one of the malformations was treated surgically. No survival of neonate with severe or critical heart defect and extracardiac malformation was observed if both ECM and CHD required surgical treatment during the first 28 days of postnatal life.

In the literature only two survivals have been reported of neonates with prenatal diagnosis of DH and CHD (Table 2, positions 4 and 9). In both cases the heart's

septal or atrial defects did not constitute indication for cardiosurgical intervention. These defects would not be considered as severe or critical from the viewpoint of prenatal cardiology.

Some cases of CHD complicated by DH might be still diagnosed postnatally but these are likely to exist in the wide spectrum of these malformations that are significantly less severe.

CONCLUSION

The presented analyses of cases from the literature, as well as those observed by our Department, confirm the unfavorable prognosis for neonates in cases of severe or critical prenatally detected congenital heart defect with coexisting diaphragmatic hernia. Additional cases with milder forms may survive neonatal surgery for diaphragmatic hernia and later on (in infancy) surgery for heart defect.

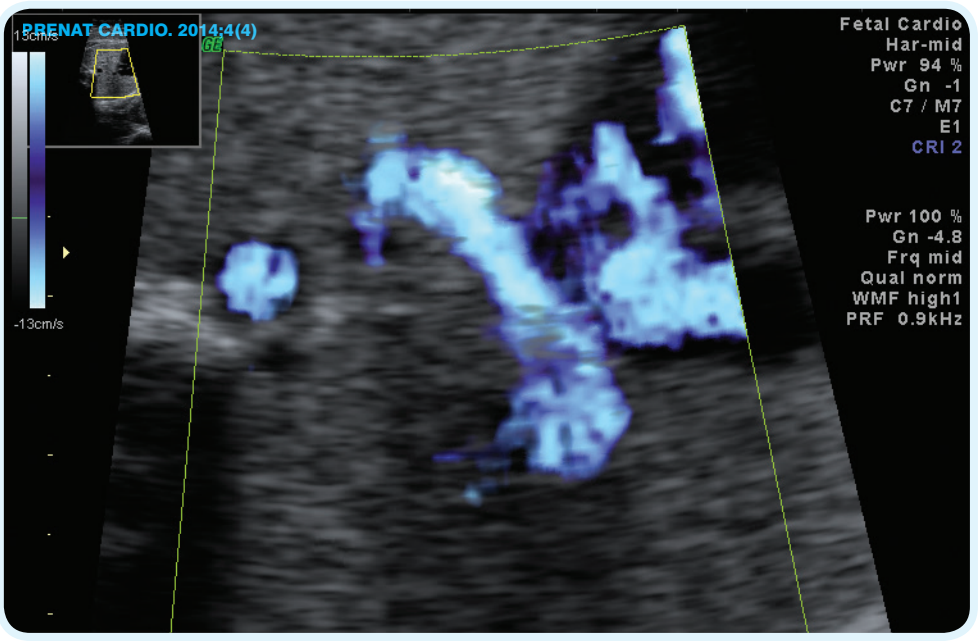


Figure 3. Abnormal pulmonary venous drainage behind the left atrium (case nr 11, table 1)

Authors and division in work:
K. Więckowska: data search and first draft
M. Słodki: discussion and correction of the manuscript
L. Dudarewicz: discussion and correction of the manuscript
H. Moczulska: data search, photo search, discussion, submission
Z. Pietrzak: discussion and correction of the manuscript
M. Respondek-Liberska: concept of the research, correction of the paper, final version
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