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CONGENITAL HEART DEFECTS COEXISTING WITH OMPHALOCELE - THE IMPORTANT PROGNOSTIC FACTOR

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Abstract

Introduction: The aim of this study was to evaluate the following parameters of fetuses and neonates with omphalocele: the prevalence of coexisting congenital heart defects (CHD), abnormalities in heart function and the impact of coexisting CHD on fetal and neonatal survival.

Material and methods: The study group consisted of 69 fetuses with omphalocele diagnosed and monitored at the Department of Prenatal Cardiology in our Institute in the years 2007-2017. The retrospective analisis of patients' data was performed. For statistical analysis we used Chi-square test, t-Student test and U Mann-Whitney test..

Results: In the studied group omphalocele was an isolated defect in 31.9% of the cases (22/69), in 68.1% (47/69) coexisting defects were present, in 49.3% (34/69) the coexisting defect was CHD. The most common CHD coexisting with omphalocele were ventricular septal defect (VSD), double outlet right ventricle (DORV) and atrio-ventricular septal defect (AVSD). Abnormalities of heart function were present in 43.5% (30/69) of fetuses with omphalocele: 23.5% (8/34) with normal heart anatomy and in 62.9% (22/35) with CHD. Statistically significant differences between the group with normal heart anatomy and the group with CHD regarded: Cardiovascular Profile Score (CVPS) (median 10 points vs median 9 points, U Mann-Whitney test p=0.034), neonatal birth weight(mean 3253 g vs median 2700 g, U Mann-Whitney test p=0.003), Apgar score (median 8 vs median 7, U Mann-Whitney test p=0.038) and survival rate until discharge from hospital (85% vs 52.9%, Chi-square test p=0.034). The comparison of data from 2007-2017 with data obtained from similar analysis performed in our center in 1999-2006, revealed significant improvement in the early detection of omphalocele (median 14.5 weeks of gestation vs mean 25.4 weeks of gestation), gestational age of delivery (mean 38 weeks of gestation vs mean 34 weeks of gestation) and survival rate until discharge both in neonates with normal heart anatomy and coexisting CHD (85% and 52.9% vs 70% and 23% respectively) .

Conclusions: 1. The presence of coexisting CHD is an important prognostic factor in fetuses and neonates with omphalocele, so early fetal echocardiography should be performed in every case of omphalocele. 2. During the last decade (2007-2017), in contrast to years 1999-2006, we observed significant improvement in early and complete prenatal diagnosis of omphalocele. 3. We observed improvement in strategy of obstetrical management resulting in delivering neonates in a more advanced gestational age both in the group with normal heart anatomy and the group with coexisting CHD.

Key words: *omphalocele, congenital heart defect, prenatal diagnosis, fetal echocardiography*

INTRODUCTION

Congenital umbilical hernia (lat. *omphalocele, exomphalos*) is an abdominal wall defect, in which organs of abdominal cavity partially or completely herniate into the umbilical cord. The sac of the hernia is composed of parietal peritoneum and amnion. Hernia usually contains liver and loops of small intestine, rarely stomach, colon or spleen[1,2]. In many cases omphalocele coexists with other congenital defects.It may also be present in syndromes of defects, such as Beckwith-Wiedemann Syndrome or Pentalogy of Cantrell[3,4]. The prevalence of congenital heart defects (CHD) coexisting with omphalocele during prenatal life and its influence on prognosis in this group of patients is not sufficiently described in the existing medical literature. The objective of this study was to perform a complete and detailed analysis of the subject. The aim of this study was to evaluate the following parameters of fetuses and neonates with omphalocele: the prevalence of coexisting congenital heart defects (CHD) and abnormalities in heart function and the impact of coexisting CHD on fetal and neonatal wellbeing and survival.

The second objective was to evaluate the differences in the diagnosis and therapy of patients with omphalocele in our center during the last two decades.

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MATERIAL AND METHODS

The study group consisted of 69 fetuses with omphalocele diagnosed and monitored at the Department of Prenatal Cardiology in our Institute between 2007-2017. The retrospective analysis of data stored in hospital databases File Maker Pro, Fetal Pathology and CliniNet was performed. The analyzed data consisted of : maternal obstetrical history (maternal age, gravidity, a number of early pregnancy losses), fetal kariotype, number of continued pregnancies, number of pregnancy terminations and intrauterine fetal demises, gestational age during detection of the defect, gestational age during the first diagnostic fetal echocardiography, prevalence and type of coexisting CHD and abnormalities of fetal heart function, prevalence and kind of coexisting extracardiac defects, organs contained in the hernia, amniotic fluid volume, prevalence of intrauterine growth restriction, data regarding perinatal period and neonatal follow-up (gestational age during delivery, way of delivery, neonates gender, neonatal birth weight, Apgar score in first minute after delivery, duration of hospital stay and survival rate until discharge from hospital.

For nominal variables, percent values were calculated. For continuous variables with normal data distribution, means and standard deviations were calculated, for continuous variables with non-normal data distribution median values were calculated. The nominal variables were compared using the Chi-square test, the continuous variables were compared using t-Student test in case of normal data distribution or using U Mann-Whitney test in case of non-normal data distribution.

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RESULTS

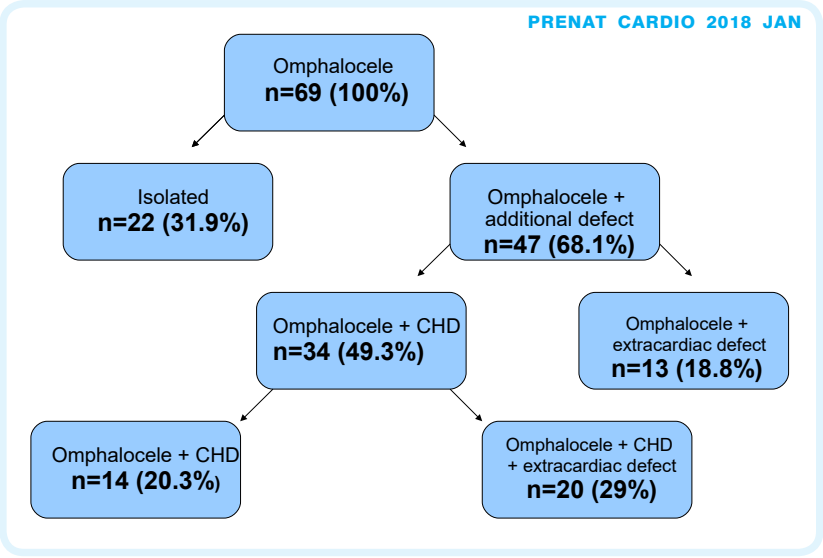
The mean age of gravida during detection of fetal omphalocele was 29.9 years (n=69, SD +/- 6.6, median 31, min. 17, max. 43). For 50% (34/68) of gravidas it was the first pregnancy, for 27.9% (19/68), the second pregnancy, for 14.7% (10/68), the third pregnancy and for 7.4% (5/68) it was more than third pregnancy. 75% (51/68) of gravidas presented unremarkable obstetrical history, 14.7% (10/68) presented medical history of 1 early pregnancy loss, 4.4% (3/68) 2 pregnancy losses, 1.5% (1/68) had 3 or more pregnancy losses. 4.4% (3/68) of gravidas presented a history of ectopic pregnancy. There were 4 twin pregnancies in the study group, 2 cases among them were conjoined twins. These pregnancies were not included in further analysis regarding perinatal period and neonatal outcome. The median gestational age of detection of omphalocele during the obstetrical ultrasound examination was 14.5 weeks of gestation (n=68, mean 16.9, SD +/- 5.5, min. 10, max. 35). The mean gestational age of first diagnostic examination in a referral center for fetal echocardiography was 23.9 weeks of gestation (n=69, SD +/- 5.9, median 24, min. 12, max. 36).

The median of time interval between detection of omphalocele by obstetrician and first diagnostic examination in a referral center was 7 weeks (n=68, mean 7.07, SD +/- 5.95, min. 0, max. 24).

The sac of the hernia contained liver in 88.2% (45/51) of cases, intestines in 45.1% (23/51) and stomach in 23.5% (12/51). In an isolated case the entrance of the hernia reached the thoracic wall and the sac of the hernia contained fetal heart.

In 25.8% (17/66) of fetuses, the anomalies of umbilical cord were present, such as the single umbilical artery, which was present in 16.7% (11/66) of cases and cysts of umbilical cord, which was present in 9.1% (6/66) of cases. In 23.2% (16/69) of cases, the ultrasound examination in referral center revealed polyhydramnion. Ascites was present in 11.6% (8/69) of the fetuses.

Intrauterine growth restriction was present in 23.2% (16/69) of fetuses. The result of karyotype examination was known for 47.8% (33/69) of the fetuses and it was normal in 90.9% (30/33) of them. There was 1 case of Down syndrome (47 XY + 21), 1 case of Edwards syndrome (47 XY + 18) and 1 case with karyotype 46,XX,der(22)t(3;22)(q23;q13). Omphalocele was an isolated defect in 31.9% (22/69) , in 68.1% (47/69), other congenital defects were present. Congenital heart defects (CHD) were diagnosed in 50.7% (35/69) of the fetuses. The postnatal echocardiographic verification



Diag.1. Prevalence of congenital heart defects and extracardiac malformations coexisting with omphalocele

of CHD revealed 2 false positive cases and 1 false negative case, giving 49.3% (34/65) neonates with CHD. The prevalence of congenital defects coexisting with omphalocele is presented in Diagram nr 1.

The most common CHD coexisting with omphalocele were ventricular septal defect (VSD) 23.5% (8/34), double outlet right ventricle (DORV) 20.6% (7/34) , atrioventricular septal defect (AVSD)- 11.8% (4/34) and ectopy of the heart 11.8% (4/35) of CHD (Table 1) .

Congenital heart defects (CHD) coexisting with omphalocele (n=34)	
Type of CHD	**n (%)**
Ventricular septal defect (VSD)	8 (23,5%)
Double outlet right ventricle (DORV)	7 (20,6%)
Atrio-ventricular septal defect (AVSD)	4 (11,8%)
Ectopia cordis	4 (11,8%)
Complex unclassified CHD	3 (8,8%)
Atrial septal defect (ASD) primum	3 (8,8%)
Tetralogy of Fallot (ToF)	2 (5,9%)
Anomalous systemic venous return	2 (5,9%)
Truncus arteriosus communis (TAC)	1 (2,9%)

Table 1.Congenital heart defects (CHD) coexisting with omphalocele (n=34)

The data of postnatal echocardiographic verification of prenatal diagnosis was available for 10 of the cases. In 40% (4/10) postnatal diagnosis was exactly the same as the prenatal diagnosis. In 40% (4/10), postnatal echocardiography confirmed the CHD, but the diagnosis was modified. In 20% (2/10), the prenatal diagnosis of CHD was false positive (1 case of VSD, and 1 case of anomalous pulmonary venous return). There was 1 false negative case of atrial septal defect type II, however this type of CHD can not be diagnosed prenatally (Table 2).

In 43.5% of the fetuses with omphalocele, an abnormal heart axis was present (70-100 degrees).

In 43.5% (30/69) abnormalities of heart function were present: they were detected in 23.5% (8/34) of the fetuses with normal heart anatomy and in 62.9% (22/35) of the fetuses with CHD.

The most common abnormalities of heart function were cardiomegaly 20.3% (14/69), tricuspid valve regurgitation 11.6% (8/69) and pericardial effusion 10.1% (7/69) (Table 3) .

The sufficiency of the fetal circulatory system was evaluated using Cardiovascular Profile Score (CVPS). The median of CVPS result for fetuses with normal heart anatomy was 10 (n=27; mean 9.74; SD+/- 0.53; min. 8, max. 10), while the median of CVPS for fetuses with CHD was 9 (n=20; mean 9; SD+/- 1.26; min. 6, max. 10). The difference was statistically significant **(U Mann-Whitney test p=0.034).**

The most common extracardiac congenital defects coexisting with omphalocele were abnormalities of the limbs (long bone shortening, club feet and deformation of hands) which were present in 17.4% (12/69), the abnormalities of the axial skeleton (hypoplastic thorax, scoliosis and deformation of ribs) which were present in 14.5% (10/69) and defects of fetal brain and cranium (anencephaly, ventriculomegaly and deformation of cranium), which were present in 13% (9/69) of the fetuses (Table 4) .

Among the 69 fetuses with omphalocele diagnosed in the prenatal period, neonatal follow-up was available for 44 of them. There was 1 case of intrauterine fetal demise. In 5 cases (11.4%), pregnancy was terminate. In all of these 5 cases additional CHD and extracardiac defects were present. In 86.4% (38/44) of cases the pregnancy resulted with live birth. 98.3% (28/30) of newborns were delivered via cesarean section. There were 2 vaginal deliveries, in one of them obstetrical forceps were used. 52.6% (20/38) of the neonates were female, 47.4% (18/38) were male. In the group of fetuses with omphalocele and normal heart

Prenatal diagnosis	Postnatal diagnosis	Similarities	Differences
Atrio-ventricular septal defect (AVSD)	Ventricular septal defect (VSD)	The ventricular septal defect was present (VSD)	There was no atrial septal defect.
Main pulmonary artery valve stenosis	Main pulmonary artery valve stenosis and ventricular septal defect (VSD)	The main pulmonary artery valve stenosis was present.	The ventricular septal defect (VSD) might have been irrelevant for fetal well-being
Double outlet right ventricle (DORV) with ventricular septal defect (VSD)	Ventricular septal defect (VSD) and atrial septal defect (ASD)	Ventricular septal defect (VSD)	The normal outlet of great arteries might have been difficult to evaluate due to elevation of fetal diaphragm caused by omphalocele and due to a VSD
Anomalous systemic venous return	Atrial septal defect		Ostium secundum atrial septal defect can not be detected by fetal echocardiography; the echocardiographic examination of the neonate did not reveal the azygos vein, which was described in the fetus
Partial anomalous pulmonary venous connection	Normal heart anatomy		False positive result
Ventricular septal defect (VSD)	Normal heart anatomy		False positive result

Table 2.Differences between prenatal and postnatal echocardiographic diagnosis

Abnormalities of fetal heart function in fetuses with omphalocele (n=69)	
Cardiomegaly	20,3% (n=14)
Tricuspid valve insufficiency	11,6% (n=8)
Pericardial effusion	10,1% (n=7)
Disproportion in favor of right side of the heart	7,2% (n=5)
Mitral valve insufficiency	7,2% (n=5)
Abnormal flow in peripheral vessels	4,3% (n=3)
Impaired myocardial contractility	2,9% (n=2)
Myocardial hypertrophy	1,4% (n=1)
Common atrio-ventricular valve insufficiency	1,4% (n=1)
Bidirectional foramen ovale flow	1,4% (n=1)

Table 3. Abnormalities of fetal heart function in fetuses with omphalocele (n=69)

anatomy, the median of gestational age during delivery was 38 weeks (n=16; mean 37.6; SD+/- 1.7; min. 33, max. 40), while in the group of fetuses with omphalocele and CHD, the median of gestational age during delivery was also 38 weeks (n=16; mean 37; SD+/- 2.9; min. 27, max. 40). There were no statistically significant difference between the groups (U Mann-Whitney test p=0.29).

The mean birth weight of neonates with omphalocele and normal heart anatomy was 3253g (n=16; SD+/-532.9g; median 3370g; min. 1940g; max. 4100g), while the median of birth weight of neonates with omphalocele and CHD was 2700g (n=17; mean 2594g; SD+/- 689.2g; min. 1300g, max. 3400 g). The observed difference was statistically significant **(U Mann-Whitney test p=0.003).** The median of Apgar score in the 1st minute of life in the group of neonates with omphalocele and normal heart anatomy was 8 (n=16; mean 7.9; SD+/-1.5; min. 6; max. 10). The median of Apgar score in the 1st minute of life in the group of neonates with omphalocele and CHD was 7 (n=16; mean 6.4; SD+/-2; min. 2; max. 9). The observed differences were statistically significant **(U Mann-Whitney test p=0.038).** In the group of surviving newborns with omphalocele and normal heart anatomy, the median duration of hospital stay was 35.5 days (n=14; mean 42.4; SD+/- 33.7; min. 6, max. 118), while that for a group of surviving newborns with omphalocele and CHD was 53 days (n=9; mean 69.3; SD+/- 60.2; min. 11, max. 210). The difference between the groups was not statistically significant **(U Mann-Whitney test p=0.28).**

The survival rate until discharge from hospital in all neonates with omphalocele was 70.3% (26/37). In the group of newborns with omphalocele and normal heart anatomy the survival rate until discharge from the hospital was 85% (17/20). In the group of newborns with omphalocele and CHD the survival rate until discharge from the hospital was 52.9% (9/17). The observed difference was statistically significant **(Chi-square test p=0.034).** The neonatal outcome of omphalocele cases was summarized in Table 5.

DISCUSSION

Omphalocele develops between 6th and 10th week of embryogenesis. In that period, a physiological herniation of the embryonic midgut into the umbilical cord is present. However, in some cases, the intestines do not move back into the abdominal cavity and other organs, most commonly the liver, may herniate into the umbilacal cord as well[5]. The prevalence of omphalocele is 2-2.5 cases per 10 000 live births. The high proportion of pregnancies with omphalocele is terminated or end with intrauterine fetal demise, therefore the prevalence of this defect in the group of fetuses may be higher than in the group of neonates[6,7,8,9]. Due to universal use of ultrasound diagnosis in modern obstetrics, omphalocele may already be detected in the Ist trimester. In our study group, the median of gestational age during detection of

Group of defects	Prevalence	Defects
Abnormalities of the limbs	17,4% (12/69)	Long bone shortening (n=5) Club feet (n=5) Deformation of hands (n=4)
Abnormalities of the axial skeleton	14,5% (10/69)	Hypoplastic thorax (n=5) Scoliosis (n=5) Deformation of ribs (n=4) Deformation of sternum (n=1)
Defects of fetal brain and cranium	13% (9/69)	Anencephaly (n=2) Deformation of cranium (n=2) Ventriculomegaly (n=2) Holoprosencephaly (n=1) Agenesis of corpus callosum (n=1)
Digestive tract defects	5,8% (4/69)	Esophageal atresia (n=2) Duodenal atresia (n=2) Anal atresia (n=2)
Defects of urinary system	4,3% (3/69)	Hydronephrosis (n=2) Polycystic kidneys (n=1)
Spina bifida	4,3% (3/69)	
Defects of reproductive organs	2,9% (2/69)	Hydrometrocolpos (n=2)
Pentalogy of Cantrell	2,9% (2/69)	
Conjoined twins	2,9% (2/69)	
Non-immune fetal hydrops	1,4% (1/69)	

Table 4. Extracardiac defects coexisting with omphalocele (n=69)

	All liveborn omphalocele cases with known follow-up (n=37)	Isolated omphalocele (n=20)	Omphalocele with CHD (n=17)	Statistical significance
Gestational age during delivery	38 weeks (median)	38 weeks (median)	38 weeks (median)	Not significant (U Mann-Whitney test p=0.29)
Birthweight	**3000 g (median)**	**3253 g (median)**	**2700 g (median)**	**Significant (U Mann-Whitney test p=0.003)**
Apgar score in 1st minute of life	**7 (median)**	**8 (median)**	**7 (median)**	**Significant (U Mann-Whitney test p=0.038)**
Duration of hospital stay (survivors only).	41.5 days (median)	35.5 days (median)	53 days (median)	Not significant (U Mann-Whitney test p=0.28)
Survival until discharge from hospital	**70.3%**	**85.0%**	**52.9%**	**Significant (Chi-square test p=0.034)**

Table 5.Outcome of neonates with omphaolcele

this defect was 14.5 weeks. It is a very big accomplishment in comparison to other European centers, where the mean gestational age of detection of omphalocele was 15-22 weeks of gestation[9,10]. The significant improvement of early detection of omphalocele in Poland during the last two decades is remarkable. The analysis performed in our center between 1999-2006 revealed, that the mean gestational age of detection of omphalocele was 25.4 weeks of gestation. In our study group, the defect was detected more than 10 weeks earlier. The early detection of omphalocele by obstetricians results in an earlier echocardiographic examination of fetal heart performed in a referral center for prenatal echocardiography. In our study group, first fetal echocardigraphy was performed at mean 23.9 weeks of gestation. In comparison, in years 1999-2006 the mean gestational age of fetal echocardiography was 27.4 weeks[11].

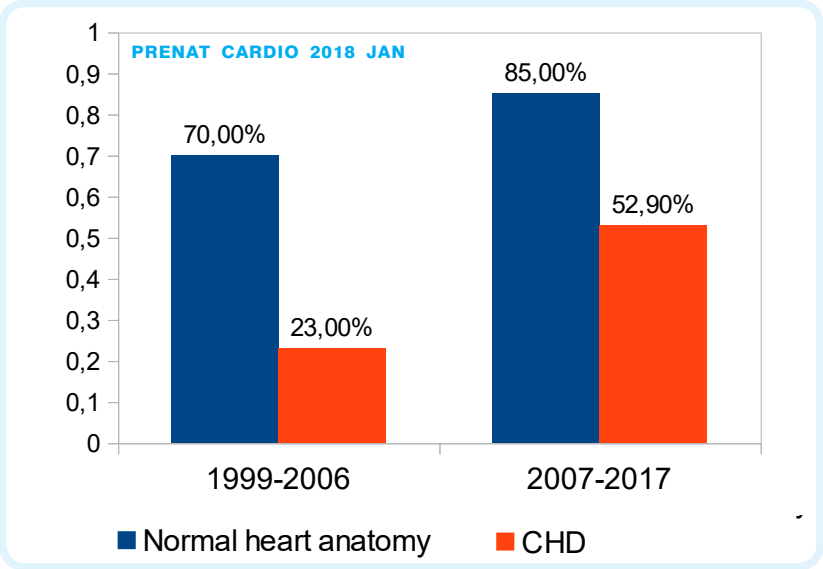
The coexistence of omphalocele with chromosome abnormalities and other genetic disorders is widely described in medical literature. Chromosome abnormalities in this group of patients occured in 18-54% of cases. The most common are trisomy of the 18th chromosome and trisomy of the 13th chromosome[7,8,9,10,12,13,14]. Unexpectedly, in our study group, chromosome abnormalities were diagnosed only in 3 of 33 (9%) fetuses. The analysis performed in our center in years 1999-2006 gave similar results, chromosomal abnormalities were detected in 2 of 12 (16.7%) of the fetuses. It is noteworthy that in the years 1999-2006, only 12 of 83 (14.5%) of the fetuses with omphalocele had karyotype examination performed, whereas in our study group karyotype was evaluated in 33 of 69 (47.8%) of the fetuses[11]. This improvement resulted from an increasing availability of invasive genetic diagnosis, greater awareness of doctors and patients regarding genetic diagnosis and earlier detection of omphalocele in obstetrical ultrasound.

In our study group, omphalocele coexisted with additional congenital defects in 68% of the cases. Our results are similar to data from medical literature, where coexisting additional defects occur in 30%-80% of the cases[7,8,12]. Our previous analysis from years 1999-2006 also provided similar results, describing additional defects in 60% of cases[11]. In our study group, the most common congenital defects coexisting with omphalocele were CHD, which were diagnosed in 49.3% of the cases. In 20.3% of the cases, CHD were the only additional defect, in 29% other extracardiac defects were also present. Our previous analysis from years 1999-2006 gave similar results: CHD was present in 44% of the cases with omphalocele. In 18% CHD was the only additional defect, in 26% other extracardiac defects were also present[11]. High prevalence of CHD in patients with omphalocele is also confirmed by the data from international medical literature, where CHD is reported in 15%-50% of the cases[5,6,7,15,16].

Boulton et al. described abnormal heart axis with an excessive leftward rotation of the apex in more than a half of fetuses with omphalocele. The abnormal axis was also common among cases with normal heart anatomy[17]. Our analysis gave corresponding results, abnormal heart axis was present in 43.5% of the cases.

In our study group abnormalities of fetal heart function were present in 43.5% . They were more common in fetuses with coexisting CHD (62.9%), but were also present in 23,5% of fetuses with normal heart anatomy. Our analysis from years 1999-2006 gave similar results: abnormalities of fetal heart function were present in 33% of the fetuses with omphalocele and normal heart anatomy[11].

The fact, that our study revealed the common coexistence of omphalocele and CHD, which is also confirmed by data from medical literature, inspired us to analyze the impact of cardiovascular abnormalities on prognosis and neonatal outcome in this group of patients. According to data from international medical literature, 33-77% of pregnancies with omphalocele are terminated[6,7,9]. Due to this, collecting a sufficiently numerous study group is hard to achieve. However, due to sociological and cultural factors in Poland, the percentage of terminated pregnancies is exceptionally low in comparison to other European countries. In our study group, 11.4% of pregnancies were terminated. The termination rate in our previous analysis from years



Diag.2. Survival rate until hospital discharge in neonates with omphalocele

1999-2006 was even lower- only 2,7%[11]. As a result, we were able to collect the unique study group of fetuses and neonates with omphalocele and coexisting CHD and analyze the follow-up.

The presence of CHD resulted in slight worsening of the sufficiency of the fetal cardiovascular system. In the group with omphalocele and normal heart anatomy the median of CVPS result was 10 points, whereas in fetuses with coexisting CHD the median of CVPS result was 9 **(U Mann- Whitney test p=0.038).** The difference was statistically significant but has not the clinical meaning. In our study group the mean gestational age of delivery was 38 weeks. It is a significant improvement in comparison to years 1999-2006, when the mean gestational age of delivery was 34 weeks[11]. The birth weight of neonates with coexisiting CHD was significantly lower (2700g) in comparison to neonates with normal heart anatomy (3253 g) **(U Mann-Whitney test p=0.003).** The Apger score, in the 1st minute after delivery, was lower in neonates with coexisting CHD. If CHD was present, the median Apgar score was 7 points, whereas in neonates with normal heart anatomy the median Apgar score was 8 points **(U Mann-Whitney test p=0.038)**. It is worth noticing, that in our study group Apgar score improved in comparison to years 1999-2006, when the mean Apgar score of neonates with omphalocele was 6 . The survival rate until discharge from hospital among all the neonates with omphalocele included in our study was 70.3%. Our results are close to the results from other centers worldwide, where the survival rate is reported to be 73%-87.5%. However, it is important to highlight that international medical literature in majority describes cases of isolated omphalocele, whereas in our study group both isolated cases and cases with other coexisiting defects were described[13,18,19].

In the group of neonates with omphalocele and normal heart anatomy the survival rate was 85%, whereas the survival rate in the group with coexisting CHD was significantly lower (52.9 %) **(Chi-square test p=0.034).**

In the years 1999-2006, the survival rate of neonates with isolated omphalocele treated in our center was 70%, whereas the survival rate of neonates with coexisting CHD was 23%[11] (Diagram 2).

The significant improvement of survival rate of neonates with omphalocele in our center in the last two decades is evident. Due to advances in prenatal diagnosis and impclose to term, leading to delivery of more mature newborns. Undoubtedly, the improvement in the field of neonatology, pediatric intensive care, pediatric surgery, pediatric cardiology and pediatric cardiac surgery was also an important factor contributing to increase of survival rate of neonates with omphalocele, both the isolated cases and cases with coexisting CHD.

CONCLUSIONS

The presence of coexisting CHD is an important prognostic factor in fetuses and neonates with omphalocele, therefore fetal echocardiography should be performed in every case of omphalocele.

During the last decade (2007-2017), in comparison to years 1999-2006, we observed significant improvement in early and complete prenatal diagnosis of omphalocele.

We observed improvement in strategy of obstetrical management resulting in delivering neonates in a more advanced gestational age both in the group with normal heart anatomy and the group with coexisting CHD.

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