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HLHS – congenital heart defect in High Risk Pregnancy Unit in 2001-2007

HLHS – wrodzona wada serca na materiale Kliniki Patologii Ciąży w latach 2001-2007

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Abstract

Background: *HLHS (hypoplastic left heart syndrome) is a serious congenital heart defect. In neonates with congenital heart disease HLHS accounts for nearly 25% of neonatal deaths.*
The etiology of HLHS is unknown. At present we expect to have those cases diagnosed in the second trimester of pregnancy.
Aim: *The main aim was to find the rate and outcome of the congenital heart defect – left heart hypoplasia (HLHS).*
Material and methods: *All cases of this heart defect found in the High Risk Pregnancy Clinic in 2001-2007 were presented. Twice the congenital malformation was diagnosed after delivery and once during pregnancy - in the second trimester. All babies were delivered between 38 and 39 weeks of pregnancy, one by the spontaneous normal delivery, one by elective caesarean section and one by emergency caesarean section because of fetal distress during the delivery. All of the newborns were transported to the Pediatric Cardio surgery Unit of M.U. and operated in first weeks after delivery.*
Results: *In two cases the operation was complicated and children died due to the cardiac arrest. One of the babies died before surgical correction. Two of the women, whose babies died after the operation gave birth to healthy newborns with no cardiac abnormalities.*
Conclusion: *The HLHS is the most serious heart congenital malformation diagnosed often after the delivery because of lack of sufficient diagnostic system in healthy women. The prognosis for the children is bad, but the chance of giving birth to healthy children in the future is good.*

Key words: **hypoplastic left heart syndrome** / **congenital heart defect** / **neonate** /

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Streszczenie

Wstęp: *HLHS (Zespół niedorozwoju lewego serca) należy do ciężkich zespołów wrodzonych wad serca. Wśród noworodków urodzonych z różnymi wadami, 25% zgonów dotyczy dzieci z tą anomalią. Etiologia zespołu HLHS jest ciągle nieznana. Obecnie spodziewamy się rozpoznania tej wady już w II trymestrze ciąży.*

Cel: *Głównym celem pracy była ocena częstości występowania zespołu HLHS oraz dalszych losów osób z tą wadą.*

Materiał i metody: *Wszystkie przypadki z rozpoznaniem tej anomalii w Klinice Patologii Ciąży z lat 2001-2007 zostały zaprezentowane. Dwukrotnie wada została rozpoznana podczas ciąży, a w jednym przypadku pourodzeniowo. Wszystkie noworodki były urodzone o czasie- między 38 a 39 tygodniem ciąży, jeden drogami natury a dwa drogą cięcia cesarskiego – jeden cięciem elektywnym a kolejny z powodu zagrożenia wewnątrzmacicznego podczas porodu. Wszystkie noworodki po porodzie przenoszone były do Kliniki Kardiochirurgii Uniwersytetu Medycznego w Łodzi, a następnie – przygotowywane do operacji w pierwszym tygodniu życia.*

Wyniki: *W dwóch przypadkach wystąpiły komplikacje pooperacyjne na kolejnych etapach złożonych operacji i dzieci zmarły na skutek nagłego zatrzymania krążenia. Jeden noworodek zmarł przed wykonaniem korekty operacyjnej. Dwie kobiety, których dzieci zmarły w trakcie kolejnych etapów korekcji chirurgicznej w 2007 roku urodziły zdrowe donoszone noworodki.*

Wnioski: *HLHS jest poważną anomalią budowy serca, która często rozpoznawana jest dopiero po urodzeniu ze względu na niedostatecznie skuteczną diagnostykę ultrasonograficzną w grupie zdrowych ciężarnych.*
Rokowanie dla dzieci jest poważne, chociaż szansa na urodzenie zdrowego dziecka w kolejnej ciąży jest duża.

Słowa kluczowe: **zespól niedokrwistości lewego serca**/ **wrodzona wada serca** / / **noworodek** /

Introduction

Early diagnosis-of hypoplastic left heart syndrome-HLHS is possible thanks to fetal echocardiography, which allows the clinician to find structural and functional abnormalities [1, 2, 3]. The final diagnosis is based on the exclusion of other structural abnormalities such as abnormal karyotyping [4]. Above data provide information on the basis of which the answer to fetal defects and prognosis can be found.

Several years ago hypoplastic left heart syndrome was considered to be a terminal condition. Normally, after the birth the closure of the ductus arteriosus is expected. In HLHS infants, there is a decreased oxygen perfusion, metabolic acidosis, circulatory insufficiency and finally, death [5]. Currently, HLHS can be detected in fetuses in the second trimester of pregnancy. The first step in the diagnostic procedure is to look for abnormalities in the scanned, two-dimensional image of the heart [1]. The general recommendation in all cases of HLHS is a three-stage Norwood procedure postnatal or if possible, heart transplantation. To stabilize circulation until the time of surgery, E1 prostaglandin is given [5, 6, 7].

The mode of delivery makes no difference in postnatal management. If the pregnant women belong to the risk group of congenital heart disease, the genetic ultrasound scan should be done [8]. Because of the impaired blood circulation associated with this condition, HLHS often leads to the death of neonates [9, 10].

The main aim of the study was to shed light on the rate of occurrence and general health implications of the left heart aplasia.

Materials and methods

The study was conducted between 2001-2007 in the High Risk Pregnancy Unit, in Lodz Medical University. Cases were neonates with HLHS, two of whom were diagnosed postnatal and one prenatal. The pregnancies were under the observation, the mode of the delivery, the neonates outcome were compared and further pregnancies were observed.

Results

The first subject was a 26 years old primiparous woman diagnosed with foetal HLHS following a normal delivery. Neither, family history of congenital diseases or specific risk factors were identified.

The delivery started spontaneously in the 39th week of gestation, ending in a natural way after 8 hours. The Apgar score after the first minute of delivery was 9. The weight of the male neonate was 3000 g. No visible congenital defects were found. Three days following delivery, symptoms of circulatory insufficiency appeared and the neonate was taken to the Pediatric Surgery unit. The baby died in the first week after the operation because of circulatory insufficiency.

The second woman, of 27, was diagnosed by US in the second trimester of pregnancy. She already had one two-year old girl without any congenital malformations. Interestingly, in third trimester of former pregnancy there was an episode of arrhythmia cordis, due to which echocardiography was performed. The results showed a normal construction of the foetal heart.

In the present pregnancy, echocardiography performed after the US revealed the HLHS. No other abnormalities of the foetal body were found in the US scan.

The delivery was done by caesarean section in the 38th week of pregnancy. The newborn male of 3600g. was born in a good condition. He was immediately taken to the Pediatric Surgery Unit and prepared for surgery.

To stabilize circulation prostaglandins were given. Five days after the delivery the first step of the Norwood procedure was performed. The resulting outcome of the surgery was satisfactory. Following the surgery the boy was taken home. Neurological assessment conducted during the first five months yielded some problems with the Central Nervous System. Magnetic resonance imaging showed problems with the central nervous system – visible as hydrocephalus – enlargement of the ventricles.

To eliminate the excess of cerebrospinal fluid neurosurgery was performed.

There were no complications during the post-operative period. However, the psycho-motor development of the boy was slower than in normal healthy children. In the 9th month the child died at home due to a sudden cardiac arrest. The autopsy revealed clotting in the pulmonary artery.

The third subject was a 26 year old primiparous woman who delivered by caesarean section. Caesarean section was done in 40 week of pregnancy, during the first stage of delivery because of PROM and intrauterine infection. The female neonate of 3700 g of weight was born in a good condition-Apgar score 9. On the second day after the delivery the symptoms of circulatory insufficiency appeared and the echocardiography was done. The diagnosis- HLHS was suspected. The prostaglandins were administrated to protect neonate from circulatory insufficiency. The neonate was taken into the Pediatric Surgery Clinic unit at the fourth day after the delivery. The baby died before the operation due to a sudden cardiac arrest.

The family history, pregnancy data and results of treatment are presented in the table (Table 1).

The first and the second of the women described in 2007 were pregnant again. Both of them were under the observation in the High Risk Pregnancy Unit from the first trimester of pregnancy. The ultrasound scan was done in the 12th and the 20th week of pregnancy and no abnormalities were found.

After 22 weeks of pregnancy echocardiography was performed and the circulatory system was described as normal. Both women gave birth to healthy newborns in the 39 week of gestation, the body mass was 3400 and 3250 grams. The newborns were under pediatric observation and control echocardiography was performed and showed no abnormalities.

Discussion

In present days congenital heart defects could be diagnosed before the delivery and treated as if needed. HLHS is connected with abnormal chromosomes often die in utero [10].

If the diagnosis was done before 22 weeks of pregnancy some of them were terminated (MPT) – for example in Helsinki University Hospital 35% of them [7]. If the couples decision was to continue the pregnancy the surgery- Norwood procedure or cardiac transplantation was proposed or postnatal hospice care [6].

Cardio-surgery is the only way to repair cardiac abnormalities. In cases of HLHS diagnosed in second trimester of pregnancy the successful 1st stage of operation after birth was about 60% [5]. In our cases the successful operations were performed in two cases(60,6%), but finally the babies died. More successful results of neonate's survival were presented by other authors [5, 11]. The mortality rate in HLHS syndrome was higher than in other heart defects [7, 9].

HLHS syndrome could be connected with abnormal karyotype and with structural anomalies from 12 to 21-28% [1, 2, 11]. In Turners syndrome about 62% cases presented cardiac abnormality, the presentation HLHS syndrome was found in 13% of fetuses [4]. In our observation one of the babies has the central nervous system defect – hydrocephalus. No data about nuchal translucency (NT) result was found in two cases; in one the result was normal. Surreus suggest that increased NT is in 62% connected with cardiac abnormalities [4].

The etiology of HLHS is unknown. One of the hypotheses is implicating an immune mechanism involving maternal antibodies produced in response to pharyngitis caused by group A beta-hemolytic streptococci (GABHS) [12].

Our results suggest that in our country the detection of fetal abnormalities system until now isn't sufficient to diagnose cardiac malformations in second trimester of pregnancy. The importance of prenatal ultrasound diagnosis of congenital heart anomalies is essential because of preparation to the correct prenatal management [1].

The arrhythmia coexists with heart defects in about 3% of cases and echocardiography should be applied in order to evaluate coexistence of heart defects and functional abnormalities [13].

Table I. Cases of HLHS in High Risk Pregnancy Unit 2002-2007.

Age of pregnant woman	Primiparous	Week of pregnancy termination	Neonatal weight	Diagnosed	Mode of the delivery	Outcome
26	+	39	3700	II-nd trimester	Caesarean section	Death-before operation
27	-	38	3600	II-nd trimester	Caesarean section	Death after the cardiac surgery
26	+	40	3000	After delivery	Normal way	Death after the cardiac surgery

Conclusion

The HLHS is very serious heart defect, even if cardio surgery was done the long term observation shows very high rate of complications and low rate of survival.

The chances of giving birth to healthy babies in next pregnancies are good.

References

1. Boldt T, Andersson S, Eronen M. Outcome of structural heart disease diagnosed in utero. *Scand Cardiovasc J.* 2002, 36, 73-79.
2. Brackley K, Kilby M, Wright J, [et al.]. Outcome after prenatal diagnosis of hypoplastic left-heart syndrome: a case series. *Lancet.* 2000, 30, 1143-1147.
3. Rychik J. Hypoplastic left heart syndrome: from in-utero diagnosis to school age. *Semin Fetal Neonatal Med.* 2005, 10, 553-566.
4. Claxon-McKinney B. Hypoplastic left heart syndrome. *Pediatr Nurs.* 2001,27, 245- 248, 251-252, 286.
5. Kawamata K, Watanabe K, Chiba Y, 9et al.]. Functional aortic stenosis diagnosed in fetal period. *Fetal Diagn Ther.* 2004, 19, 106-110.
6. Munn M, Brumfield C, Lau Y, [et al.]. Prenatally diagnosed hypoplastic left heart syndrome--outcomes after postnatal surgery. *J Matern Fetal Med.* 1999, 8, 147-150.
7. Mandorla S, Narducci P, Migliozzi L, [et al.]. Fetal echocardiography. Prenatal diagnosis of hypoplastic left heart syndrome. *G Ital Cardiol.* 1984, 14, 517-520.
8. Surerus E, Huggon I, Allan L. Turner's syndrome in fetal life. *Ultrasound Obstet Gynecol.* 2003, 22, 264-267.
9. Taketazu M, Barrea C, Smallhorn J, [et al.]. Intrauterine pulmonary venous flow and restrictive foramen ovale in fetal hypoplastic left heart syndrome. *J Am Coll Cardiol.* 2004, 43, 1902-1907.
10. Allen R, Benson C, Haug L. Pregnancy outcome of fetuses with a diagnosis of hypoplastic left ventricle on prenatal sonography. *J Ultrasound Med.* 2005, 24, 1199-1203.
11. Włoch A, Włoch S, Sikora J, [i wsp.]. Zaburzenia rytmu w okresie prenatalnym a stan noworodka – analiza materiału własnego. *Ginekol Pol.* 2001, 72, 968-971.
12. Verheijen P, Lisowski L, Plantinga R, [et al.]. Prenatal diagnosis of the fetus with hypoplastic left heart syndrome management and outcome. *Herz.* 2003, 28, 250-256.
13. Eghtesady P. Hypoplastic left heart syndrome: Rheumatic heart disease of the fetus? *Med Hypotheses.* 2006, 66, 554-565.