HYPOPLASTIC LEFT HEART SYNDROME AT THE TERTIARY FETAL CARDIAC CENTER: AS PLANNED, URGENT OR SEVEREST CONGENITAL HEART DISEASE? PRENATAL CLASSIFICATION FOR OBSTETRICIANS AND NEONATOLOGISTS

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Abstract

Hypoplastic left heart syndrome (HLHS) is one of the commonest heart defects detected prenatally in the world. For many years now, it has been at the very top of the list of the commonest foetal heart defects in the Polish National Registry for Foetal Cardiac Pathology (www.orpkp.pl). According to a new classification of foetal heart defects, HLHS – as an isolated heart defect – can be classified into the following three groups: severest heart defects (despite immediate surgical intervention made just after birth, nearly 100% of the infants die); severe heart defects requiring immediate cardiac intervention in a hemodynamics room; and severe heart defects requiring no immediate cardiac intervention (infants are born in a good condition of health and can be prepared for the first stage of their cardiac operation as planned). The present study looks at three cases of HLHS classified into three different groups of the new classification of foetal heart defects. In terms of specialist medical literature written to date, this classification of foetal heart defects from the point of view of prenatal hemodynamics is a novelty; it may help obstetricians and neonatologists working at referral centres to act properly at labour wards.

Key words: *HLHS, hypoplastic left heart syndrome, planned severe congenital heart disease, urgent severe congenital heart disease, the severest congenital heart disease*

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is one of the most common prenatally detected heart defects in the world. For many years now, it has been at the very top of the list of the most prevalent fetal heart defects in the Polish National Registry for Fetal Cardiac Pathology (www.orpkp.pl). According to the new division of fetal heart defects, HLHS – as an isolated heart defect – can be classified into the following three groups: severest heart defects (despite immediate surgical intervention after birth, nearly 100% of infants die); severe heart defects requiring immediate cardiac intervention in a hemodynamics laboratory; and severe heart defects requiring no immediate cardiac intervention (infants are born in good condition and can be prepared for the first stage of their cardiac operation as planned).

The present study looks at three cases of HLHS classified into three different groups using the new division of fetal heart defects. In terms of specialist medical literature written to date, this classification of fetal heart defects from the point of view of prenatal hemodynamics is a novelty; it may help obstetricians and neonatologists working at referral centers to act properly at labour wards.

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Case 1

Prima gravida aged 21, low-risk pregnancy. The results of the ultrasound examination which she underwent in the 14th week of pregnancy were normal. Nuchal translucency was 1.2 mm, and the results of her serum biochemical screening tests were normal. At 21 weeks of gestation, HLHS was diagnosed. The first consultative examination at the Prenatal Cardiology Unit was conducted at 28 weeks of gestation. In the meantime, the gravida consulted specialists at a type A referral centre. The diagnosis of HLHS was confirmed (hypoplastic, hardly movable mitral valve, atretic aortic valve, contracted left ventricle diameter with some features of fibroelastosis, apex cordis formed by the right ventricle). The function

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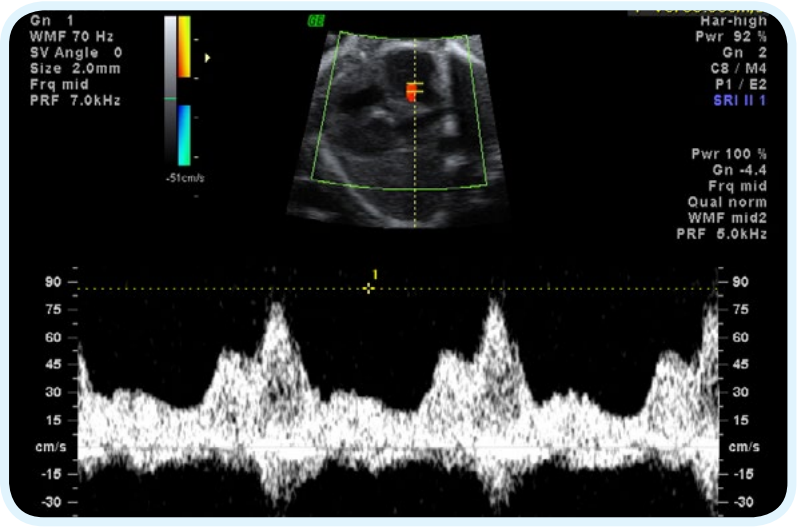
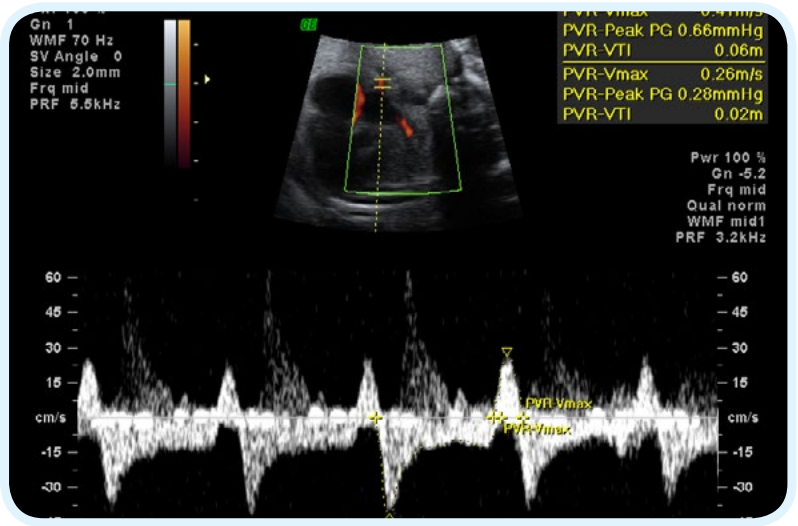
of the foramen ovale and pulmonary venous flow were evaluated by means of an echocardiogram test. The test showed a patent foramen ovale (picture 1) and normal pulmonary venous flow spectrum (picture 2). The subsequent echocardiogram test performed in the 33rd week of pregnancy produced similar results. The last prenatal echocardiogram test was performed in the 39th week of pregnancy, 24 hours before birth. The width of the foramen ovale was 4 mm, its motion was plane, and the maximum velocity of the blood flow from the left atrium to the right one was 80 cm/s. The maximum velocities of the pulmonary venous flows were 45 cm/s, 35 cm/s and 15 cm/s, with no backflow. In this case, as the postnatal health condition of the patient was predicted to be good, the heart defect was classified as a severe defect requiring no immediate cardiac intervention. The pregnant woman delivered spontaneously; the female infant weighed 3440 grams and her Apgar score was 9. The infant was given Prostin from the very first hours of life. As planned, she was transferred from the Neonatology Clinic to the Cardiology Clinic on the seventh day of life and scheduled for the Norwood procedure on the tenth day of life. Following a classical sternotomy, open-heart surgery was performed at the Cardiac Surgery Clinic. On the third day after the surgery, the patient was respiratorily efficient and, initially, fed intravenously and then orally. She was discharged in the period of body weight gain, with saturation levels of 85-89% on the 24th day following surgery. The girl is now 8 months old and has undergone the second phase of the HLHS treatment. Her overall condition is currently good.



Fot. 1. Prawidłowy szeroki otwór owalny u płodu z HLHS w 30 tyg. ciąży.
Fot. 1. Normal foramen ovale in fetus with HLHS at 28th week of gestation.



Fot. 2. Prawidłowe spektrum przepływu przez żyły płucne u płodu z HLHS w 28 tyg. ciąży.
Fot. 2. Normal pulmonary vein flow in fetus with HLHS at 28 week of gestation.

Case 2

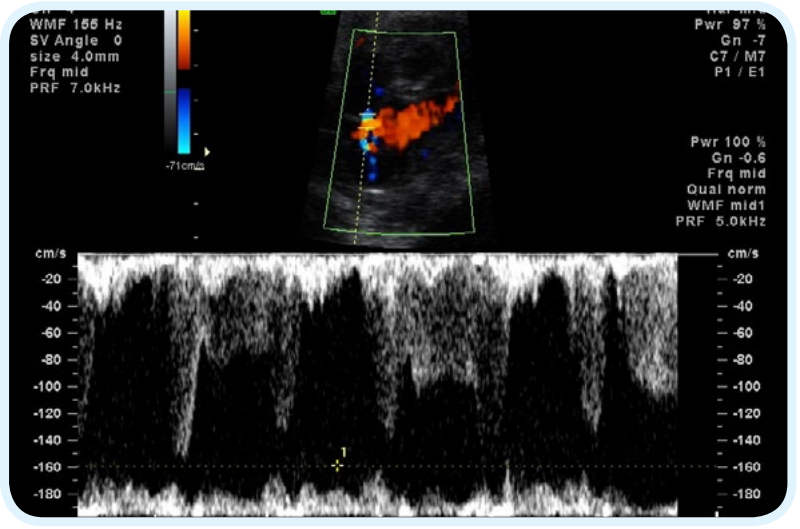
Patient aged 36, pregnant for the second time. First pregnancy resulted in a healthy son. The second pregnancy was high-risk because of the mother's age. The results of the obstetric ultrasound examination which she underwent at 13 weeks of gestation were normal. Nuchal translucency was 1.5 mm, and the results of her biochemical tests were negative. The pregnant woman decided on amniocentesis at 16 weeks of gestation, the result being 46,XY. The results of the examination which she underwent at 21 weeks of gestation were normal. The first fetal heart abnormalities were detected in the 34th week of pregnancy and the mother was referred to a prenatal cardiology centre. The diagnosis of HLHS was confirmed in the 35th week of pregnancy (the anatomical variant being similar to that in case 1). A postnatal consultative examination conducted at the prenatal cardiology centre (type C) at 38 weeks showed a 2 millimeter wide restrictive foramen ovale with accelerated left-to-right shunt with the maximum velocity of 159cm/s (picture 3), abnormal diphase pulmonary venous flow with the maximum velocity of 40 cm/s, and backflow with the maximum velocity of 30 cm/s (picture 4). The anatomical structures were described and it was pointed out that there was a foramen ovale restriction, which suggested the need for an immediate Rashkind atrial septosomy after birth. In this case, the heart defect was classified as a severe requiring immediate cardiac intervention. The results of the examination were submitted to the head of the Pediatric Cardiology Clinic. The gravida delivered spontaneously 10 days after the last echocardiogram test. Within the first 24 hours of life, the infant was administered Prostin intravenously, and underwent an immediate Rashkind atrial septosomy. Cardiac surgery was performed on the 19th day of life at the Cardiac Surgery Clinic of the Polish Mother's Memorial Hospital in Łódź. After the surgery, there were problems with weaning the infant of the ventilator. On account of the character of the infection, antibiotics were administered. On

the 28th day after the first stage of the Norwood procedure, i.e. on the 47th day of life, the male infant was discharged, with saturation levels at 80%.

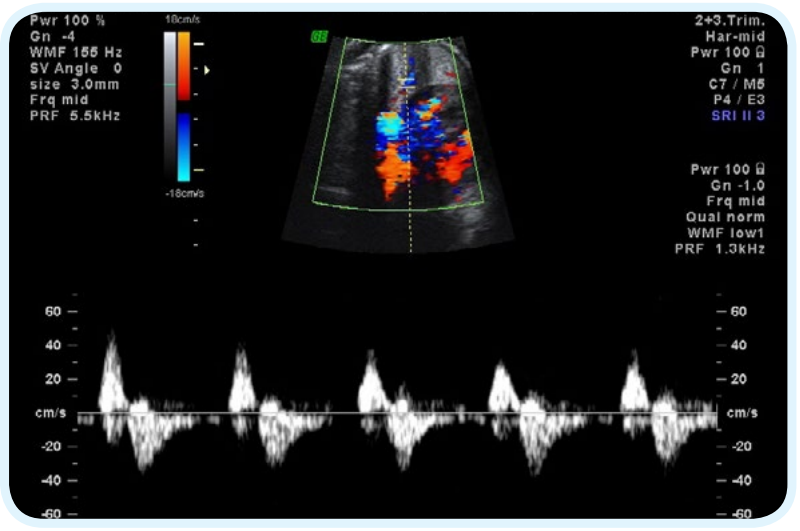
Case 3

Patient aged 38, pregnant for the third time. This pregnancy was high-risk due to the mother's age. First and second pregnancies resulted in two healthy children. The ultrasound examination which she underwent at 13 weeks of gestation showed nuchal translucency of 2.7 mm; the results of her biochemical tests were negative, and she decided against amniocentesis. The obstetric ultrasound examination which she underwent at 22 weeks of gestation revealed certain abnormalities: suspected HLHS. The first examination at a prenatal cardiology referral centre was conducted at 24 weeks of gestation; it confirmed the presence of a structural heart defect in the form of HLHS (anatomical description similar to details presented in case 1). However, power angiography showed no detectable flow through the foramen ovale. Subsequent examinations were performed at 26, 28 and 33 weeks of gestation, monitoring normal fetal biometry, normal flows in the umbilical cord vessels and ductus venous. From 33 weeks of gestation onwards, extremely abnormal pulmonary venous flows were detected. The last echocardiogram test was performed in the 38th week of pregnancy; it was supplemented with a fetal oxygen test, whose results were negative.

It was concluded that the prognosis was grave as the fetus had HLHS with a closed foramen ovale. The infant was expected to die soon after birth as the heart defect was classified as severest. The infant was born three days after the last examination; the pregnancy was completed by elective Caesarean section. The infant weighed 3031 grams and the Apgar score was 8. In the fourth hour of life, the infant was diagnosed with respiratory insufficiency and died 12 hours later.



Fot. 3. Restrykcyjny otwór owalny u płodu z HLHS w obrazowaniu dopplerowskim.
Fot. 3. Doppler image of restrictive foramen ovale in fetus with HLHS.



Fot. 4. Nieprawidłowe spektrum przepływu przez żyły u płucne u płodu z HLHS i restrykcyjnym otworem owalnym.
Fot. 4. Abnormal pulmonary vein flow in fetus with HLHS and restrictive foramen ovale.

DISCUSSION

Infants suffering from isolated congenital heart defects may need to be treated in various ways; they may require planned cardiac surgical treatments in the first weeks of life (severe heart defects requiring no immediate cardiac intervention) or cardiac interventions on the first days – or even in the first hours – of life (severe heart defects requiring immediate cardiac intervention). However, modern medicine sometimes fails to save infants' lives despite accurate prenatal prognoses and immediate surgical intervention (severest heart defects). All three cases may occur at labour wards when infants suffering from HLHS are born.

Case 1 illustrates the most frequent case in which an infant suffering from HLHS is born with a wide foramen ovale, and is given Prostin as planned in order to support the patency of the ductus arteriosus, and scheduled for the Norwood procedure (usually in the second week of life). Data collected by the Department for Diagnoses and Prevention of Congenital Malformations shows that this type of heart defect constituted about 87% of the 75 cases of HLHS examined at this unit between 2009 and 2011.

In case 2, HLHS required immediate cardiac intervention; the Rashkind atrial septosomy was performed on the first day of life. Between 2009 and 2011, such cases constituted about 13% (10 out of 75). HLHS with a continuous or restrictive interatrial septum results in the greatest number of postnatal deaths. This is why such forms of HLHS represent the most difficult challenge for prenatal diagnosis and postnatal treatment specialists. What we already know is that during pregnancy a wide foramen ovale may

Echocardiografic features	*Case 1*	*Case 2*	*Case 3*
Hypoplastic, hardly movable mitral valve	(+)	(+)	(+)
Atretic aortic valve	(+)	(+)	(+)
Contracted left ventricle diameter with some features of fibroelastosis	(+)	(+)	(+)
Apex cordis formed by the right ventricle	(+)	(+)	(+)
Bidirectional blood flow in the superior mediastinum	(+)	(+)	(+)
Foramen ovale	*Wide* >2mm, Vmax<100cm/s	*Restrictive* <2mm, Vmax >100cm/s	*Closed*
Pulmonary venous flow spectrum	*Normal*	*Abnormal*	*Totaly abnormal*

Table 1. HLHS as a defect in the three phases – similarities and differences

become more and more restricted and eventually close completely, resulting in the severest form of HLHS (case 3).

Attempts are currently being made to elaborate on the number of parameters regarding the foramen ovale function. As far as HLHS is concerned, the most extensive research has been conducted in Boston, where pulmonary venous flow spectrum is believed to be the major parameter in terms of predicting newborn infants' physical status. While evaluating the pulmonary venous flow spectrum, the following were measured: the velocity of S wave, the velocity of D wave, the velocity ratio of S wave to D wave, the velocity of A wave, the duration of A wave, the velocity time integral of A wave, the velocity time integral of the positive flow, and the ratio of the velocity time integral of the positive flow to the velocity time integral of A wave. The exhaustive research indicates that the most reliable parameter is the last one.

Another study on the prenatal diagnosis of HLHS, consisting of 39 fetuses with HLHS, verified previous reports. It is suggested in the study that all the fetuses should routinely undergo echocardiogram tests performed every four weeks on account of the possible development of their heart defects. The fetuses diagnosed with the >5 ratio of positive flow VTI to wave VTI were classified into the low-risk group for immediate postnatal cardiac intervention (i.e. severe heart defects requiring no immediate cardiac intervention in our terminology). The fetuses whose index was between 3 and 5 were classified into the at-risk group for immediate postnatal cardiac intervention, whereas those whose index was <3 were classified into the high-risk group for immediate postnatal cardiac intervention, including an optional intrauterine intervention. In our terminology, HLHS with indexes <5 are classified as severe heart defects requiring immediate cardiac intervention.

In the severest cases, i.e. prenatal closure of the foramen ovale, attempts have also been made – in several cardiology centres in the world – to broaden the foramen ovale prenatally. However, none of the attempts have been successful so far. In Poland, some attempts are being made in terms of aortic valve cardioplasty in the case of heart defects which may develop into HLHS.

Listed in Table 1, the cases presented in this study show that HLHS is a heart defect which not only needs to be detected during pregnancy, but also requires constant echocardiogram monitoring. What besets prenatal cardiologists are the following problems: the proper evaluation and prediction of neonate's physical status; the recognition of the potential need for in utero treatment; and the organisation of a team of specialists who will attempt surgical intervention in the hemodynamics laboratory.

The proposed division of heart defects, based on the three presented cases is mainly intended for obstetricians and neonatologists, but it may also make the methods adopted by the teams of specialists more efficient in the future.

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