SIGNIFICANCE OF HYPOPLAASTIC LEFT HEART SYNDROME PRENATAL DIAGNOSIS FOR OUTCOMES

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ABSTRACT

OBJECTIVE — Prenatal echocardiography is very important for diagnosis of hypoplaastic left heart syndrome, the management of affected fetuses, including parental counseling for the therapeutic options, the planning of the delivery and the postnatal care.
METHODS — Multiple B-scan planes, Doppler color flow mapping and pulsed Doppler, 3–4 Dimensional Fetal Echocardiography. Methods of the echocardiographic identification of fetal CHD are: postnatal echocardiography, angiography, surgery, or autopsy.
RESULTS AND CONCLUSION — A total of 310 fetuses which HLHS were obtained during an period between 2011–2013 years from which 161 with a prenatal diagnosis of HLHS and 149 with postnatal diagnosis were enrolled. Hypoplastic left heart syndrome can be easily recognized on prenatal ultrasound and is one of the most common serious cardiac defects diagnosed prenatally. Hypoplastic left heart syndrome (HLHS) is frequently diagnosed prenatally, but Multiple-stage palliation surgery, postoperative complications determine low quality of life as the consequence.

KEYWORDS — Fetal echocardiography, congenital heart diseases, postnatal care.

OBJECTIVE

Hypoplastic Left heart syndrome is a congenital heart disease with a more dramatic change in diagnostic approach, management and outcome. During this time , survival to the age of 5 years (including Fontan) has ranged from 50% to 69% [1].

The stages of care of HLHS are:

— pre-Stage I: fetal and neonatal assessment and management;
— Stage I: perioperative care, interstage monitoring, management strategies;
— Stage II: surgeries;
— Stage III: Fontan surgery;
— Stage VI: long-term follow-up.

The of reportes have concluded that mortalyity is not reduced if a prenatal diagnosis is made [2, 3], though some have reported improved survival [4]. At present time significance of hypoplaastic left heart syndrome prenatal diagnosis for outcoms also is permission potencial fetal intervention. In select cases? Prenatal balloon dilatation of the aortic valve has been associated with decreased progression of the left ventricular hypoplasia [5].

METHODS

Definition of fetal CHD was attempted from multiple scan planes including four-chamber, long- and short-axis, aortic arch and ductal arch views [fig. 1, 2]. We use 3–4 Dimensional Fetal Echocardiograms dimensional echo.

Doppler color flow mapping and pulsed Doppler interrogation were used to facilitate identification of great vessel relationship, location and severity of ventricular outflow obstruction [fig. 3].

Initial fetal echocardiograms were obtained between12 and 39 weeks of gestation (median 24.5 weeks).

HLHS usually are diagnosed during the first echo.

Age at first examination ranged between 17 and 41 years v (median 30 years). The median number of echocardiographic studies performed was one, ranging from one to four examinations.

We think, major cardiac malformations should be followed serially by fetal echocardiography as progressive alterations in flow may affect growth of cardiac

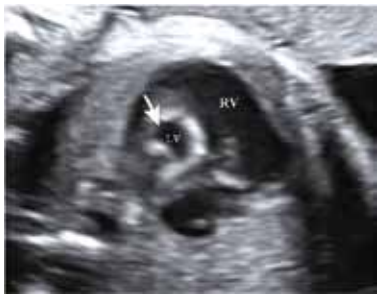

Fig. 1. *24 weeks of gestation. Hypoplastic left heart syndrome. RV — right ventricule, LV — left ventricule*

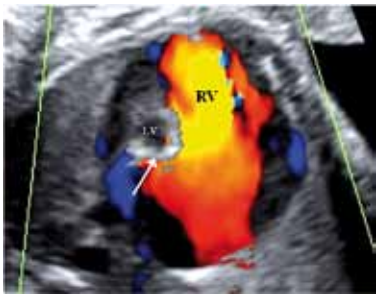

Fig. 2. *32 weeks of gestation. Hypoplastic left heart syndrome. RV — right ventricule, LV — left ventricule. Color Doppler Mapping*

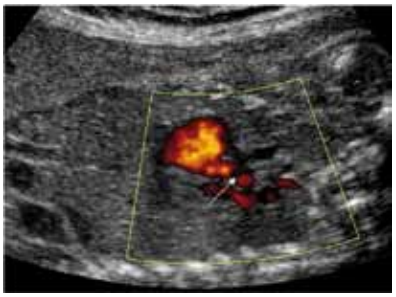

Fig. 3. *18 weeks of gestation. Hypoplastic left heart syndrome. Power Doppler Mapping. Arrow — hypoplastic aorta ascendens*

structures over time: for example, very often, after prenatal diagnosis of hypoplastic left-heart syndrome couples have been offered termination of pregnancy. But termination of pregnancy should not be proposed when it is only a small left ventricle (on echo), because many of those patients end up with only coarctation of the aorta." A second echo should be carried out in these cases.

Maternal age was from 17 to 41 years old.

— 23% of fetal echocardiograms were obtained before 18 weeks of gestation.
— 69% of fetal echocardiograms were obtained between 18–28 weeks of gestation.
— 8% of fetal echocardiograms were obtained between 29–39 weeks of gestation.
— All infants with HLHS are borne by mothers with no known risk factors.

Methods of the echocardiographic identification of fetal CHD are: Postnatal echocardiography, angiography, surgery, or autopsy.

Fetal echocardiography has opportunity to study the most important parameters of fetal heart with HLHS for postnatal surgical repair, such as left/right ventricular diastolic dimensions in M-mode, B-mode (right–to–left ventricular disproportion, ultrasound indication of endocardial fibroelastosis). Mitral valve anomaly (congenital Parachute mitral valve, stenosis/atresia), aortic valve or aortic root disease, left ventricular outflow obstruction, dimension of foramen ovale and ductus arteriosus

RESULTS AND DISCUSSION

A total of 310 fetuses which HLHS were obtained during an period between 2011–2013 years from which 161 with a prenatal diagnosis of ***HLHS and 149 with postnatal diagnosis*** were enrolled. Nobody had major extracardiac malformations and/or chromosomal abnormalities.

In our group, there are patients with HLHS, in our group, who are diagnosed prenatally and postnatally have improved survival after first-stage palliative surgery compared with those who have survived after Norwood procedure. We were not included patients after Norwood procedure in further analysis. Outcome showed at the tables 1, 2.

We don't mean the comparison of features of clinical treatment in these two groups as a We would like to analyse parameters which improve the initial clinical status of patients with HLHS.

Our results showed The Key parameter defining influence of prenatal diagnostics to surgical treatment patients with HLHS is time of arrival of the newborn in a cardiovascularsurgery hospital. Age At admission was more uniform and significantly younger in the prenatal group, than in the postnatal group. The difference follows from this indicator in time of carrying out of surgical operation. About 1 day from the moment of the diagnosis statement. There are no preoperatively metabolic acidosis, dysfunction RV in the prenatal group of patients. Dysfunction of RV takes place only in the postnatal group of patients in low interest, children with late arrival to clinic, more than 6–7 days. Children from all regions of our country come to our center, that's why, unfortunately, there may be cases of late initial diagnosis. There is significantly lower incidence of preoperative inotropic medications in the prenatal group of patients. 15–20% versus 55–60%. And significantly lower incidence of hospital lethality after the first stage as the consequence (7,1% in the prenatal group versus19,2% in postnatal group).

The reasons of lethality are various. First of all is the severity of disease. Our study demonstrates that prenatal diagnosis of HLHS was associated with improved preoperative clinical status and with improved

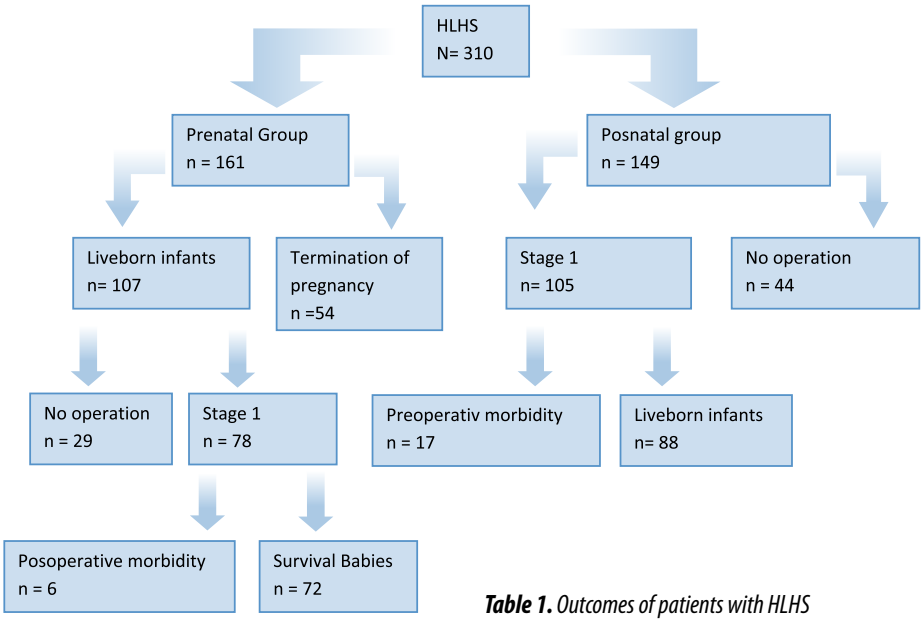

Table 1. *Outcomes of patients with HLHS*

Table 2. *Camparison of characteristics of patients with hlhs in the prenatal and postnatal groups*

PARAMETERS	Pranatal Group (n = 78)	Posnatal Group (n = 105)	P
Age at admission (hours)	5,2 ± 1,6	15 ± 7,4	p < 0,001
Age between delivery and operation (hours)	22,4 ± 3,1	34 ± 3,9	p < 0,001
preoperative Metabolic acidosis	3,5%	45%	p < 0,001
Preoperative inotropic medications	16,8%	58,4%	p < 0,001
dysfunction of RV	0,9%	4,9%	NS
Postoperative mortality	7,1%	19,2%	p < 0,05

survival after first-stage palliation in comparison with patients diagnosed after birth.

Hypoplastic left heart syndrome can be easily recognized on prenatal ultrasound and is one of the most common serious cardiac defects diagnosed prenatally. The standard "4-chamber cardiac view" used by obstetricians for screening of congenital heart disease demonstrates either a small left side or an echogenic left ventricle from endocardial fibroelastosis.

Hypoplastic left heart syndrome (HLHS) is frequently diagnosed prenatally, but Multiple-stage palliation surgery, postoperative complications determine low quality of life as the consequence.

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