Syndrome of hypoplastic left heart: a case report

I. Felesakis, E. Kontomanolis, P. Dalla, A. Liberis, G. Galazios, V. Liberis

Department of Obstetrics & Gynecology, Democritus University, Alexandroupolis (Greece)

Summary

A 36-year-old woman, gravida 5, para 3, was referred to this outpatient clinic in the 18th week of gestation for monitoring of her pregnancy. A 22-week anomaly scan confirmed hypoplastic left heart syndrome (HLHS) but the parents declined any medical intervention. Here the authors present the management of a pregnancy and the expected quality of neonatal life.

Key words: Aortic atresia; Aortic hypoplasia; Congenital heart disease; Hypoplastic left heart syndrome; Mitral stenosis; Nuchal fold.

Introduction

Hypoplastic left heart syndrome (HLHS), accounts for four percent of all cases of congenital heart disease (CHD). Infants will die soon after their birth unless they undergo surgical intervention. HLHS is the most severe form of the obstructive lesions of the left heart side.

Case Report

In the present case, a 36-year-old woman, gravida 5, para 3, of Muslim descent, rhesus positive, was referred to this outpatient clinic in the 18th week of gestation for monitoring of her pregnancy. Her obstetric history reported two labors, one preterm labor which died soon after the birth and one intrauterine death at the third trimester without a known cause, all delivered with cesarian section. Prenatal diagnosis revealed nothing abnormal. Fetal growth was assessed every two weeks. Cervical length was measured with ultrasound to estimate the risk for spontaneous delivery and performed vaginal swabs for *mycoplasma hominis, gardenella vaginalis, chlamydia trachomatis,* and *escherichia coli.* Urine cultures were collected on a monthly basis. She was also tested for mutations in the factor V G1691A (leiden), factor II (prothrombin), MTHFR genes, PAI-1 (plasminogen activator inhibitor), and gestational diabetes. Hematological and fetal growth parameters were closely monitored on an outpatient basis. Pregnancy was uncomplicated until her 22-week anomaly scan.

The 22-week anomaly scan showed an increased nuchal fold (NF = 7.92) (Figure 1). Fetal echocardiography revealed severe left ventricular hypoplasia (Figure 2). Additional cardiac anomalies included severe left atrial hypoplasia, severe left ventricular hypoplasia (the right ventricle constitutes the cardiac apex giving rise to the term "apex-forming ventricle"), interventricular communication; flow across the foramen ovale was from left to right, mitral valve hypoplasia, tricuspid valve hypoplasia, mitral valve stenosis, mild tricuspid valve insufficiency with no regurgitation, ascending aortic hypoplasia, main pulmonary artery dilatation, reversed aortic arch, and mild pericardial effusion were found. This unborn child suffered from a rare and lethal disorder. Newborns are significantly affected by this condition; their quality of life may not be so promising. It is very rare for infants with HLHS to survive at long term. A series of operations can be suggested and conducted with a limited success. Amniocentesis and termination of pregnancy were offered because of the major heart problems and the significant neonatal mortality. The parents declined because of their religious beliefs and continued the pregnancy. In 26th week of pregnancy the patient required hospitalization due to massive vaginal bleeding and negative fetal heart rate. Delivery was achieved by cesarian section.

Discussion

HLHS, a collection of anomalies, accounts for four percent of all cases of CHD. Infants will die soon after their birth unless they undergo surgical intervention. HLHS is the most severe form of the obstructive lesions of the left heart side. The way HLHS is inherited is not clearly defined. Although there is male predominance, in this case report the affected fetus is of female gender. Tricuspid regurgitation is present in more than 50% of patients with HLHS preoperatively. In the present fetus, no regurgitation was detected.

A four-chamber view with a small left ventricle and a hypoplastic ascending aorta is the common view that guides the sonographer to the diagnosis of HLHS [1, 2]. Occasionally the left ventricle can be smaller than the right one but not enough to be clearly demonstrated in the four-chamber view until the late second- or early third-trimester of pregnancy. The right ventricle becomes more spherical [3]. The left ventricular wall may show decreased contractility and appear echogenic. The presence of atrial septal restriction should be taken into consideration as it increases mortality [4]. HLHS is associated with other heart anomalies such as ventricular septal defect, aortic arch interruption, transposition of the great vessels, and central nervous system abnormalities such as microcephaly, holoprosencephaly, and agenesis of corpus callosum. Magnetic resonance imaging (MRI) testing during intrauterine life has failed to detect fetuses with possible neurodevelopmental abnormalities after birth [5]. The fetus should go through a detailed sonography to exclude sonographic markers for trisomies and Turner Syndrome and amniocentesis should be offered if they are present [6]. In Doppler real time examination, there is retrograde blood flow to ascending aorta and aortic arch.

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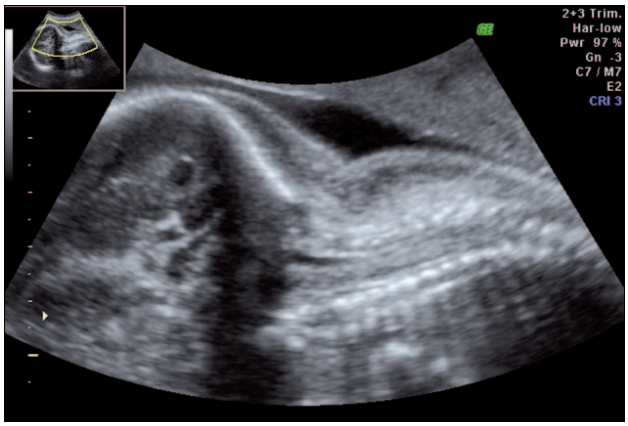

Figure 1. — Embryo's nuchal region.

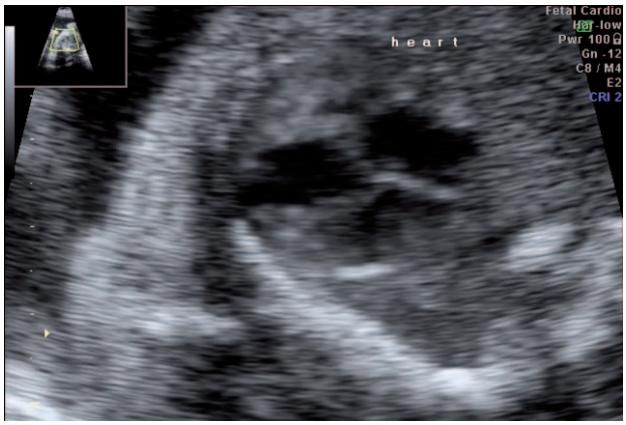

Figure 2. — Hypoplasia of the left heart.

Embryos with HLHS have a normal intrauterine growth, but their prognosis after birth is extremely poor.

If diagnosis is done before 24 weeks, termination of the pregnancy should be offered after informing the couple of the neonatal mortality and the complications of the surgical intervention. Parents need to be informed on new available intrauterine surgical interventions [7]. If expectant management is desired from the parents, the embryo should undergo series of sonographic exams. Fetal growth should be assessed on a regular basis [1].

Delivery should be performed in a tertiary center with pediatric specialists and referral to cardiothoracic surgeons for initial palliation.

Prostaglandin therapy is given to affected infants after birth for keeping ductal patency with poor outcomes, as they begin to develop heart failure in the first day. Despite improved surgical outcomes, the majority of infants continue to receive no surgical care [8]. Surgical intervention can be heart transplantation or Norwood repair, a three stage surgery [9]. Stage 1 involves anastomosis of the pulmonary artery to the aortic arch for systemic outflow, placement of a systemic-to-pulmonary arterial shunt to provide pulmonary blood flow, and arterial septectomy to ensure unobstructed pulmonary venous return; the survival rate of fetuses diagnosed in utero is in the region of 40%. Stage 2 (in the sixth month of life) involves anastomosis of the superior vena cava to the pulmonary arteries [2]. The final stage is the Fontan operation, which can be performed between the ages of 18 months and four years. The operative mortality of the Norwood operation is ten percent, the postoperative long-term effects of surgical intervention survivors may include neurodevelopmental abnormalities [9, 10].

Conclusion

HLHS is the most common heart disease. It is detectable during pregnancy and the establishment of the diagnosis compels parents to face the dilemma of discontinuing the pregnancy or having conservative management of it.

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Address reprint requests to:
I. FELESAKIS, M.D.
9, Simeonidi str.
PC54352, Pilaia (Greece)
e-mail: i.felesakis@hotmail.com