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REVIEW ARTICLE

Severe Congenital Diaphragmatic Hernia—Exemplary Management in Poland: Fetoscopic Endotracheal Occlusion

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ABSTRACT

Congenital diaphragmatic hernia (CDH) is a developmental closure defect resulting in discontinuity of the diaphragm. Abdominal viscera herniate into the chest leading to mediastinal shift and lung compression. Congenital diaphragmatic hernia occurs in approximately 1 in 4,000 live births. Vast majority of cases is on the left side of the diaphragm. Congenital diaphragmatic hernia is also associated with severe pulmonary hypoplasia and pulmonary arterial hypertension. Approximately 50 to 70% of cases of CDH are isolated, but may also be associated with rare and severe genetic syndromes. It is one of the most severe birth defects with extremely high neonatal mortality and morbidity. Prognosis is worse in cases of an abnormal chromosomal microarray, severe associated anomalies, right-sided defect, liver herniation, and lower fetal lung volume. In some countries, fetoscopic endotracheal occlusion (FETO) for severe CDH is offered to selected group of patients. Since 2014, this procedure is also available in Poland in the 1st Department of Obstetrics and Gynecology of Medical University of Warsaw. This article reviews management in diaphragmatic hernia and presents protocol implemented in our center.

Keywords: Congenital diaphragmatic hernia, Endotracheal occlusion, Fetoscopy, Tracheal occlusion.

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INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a group of closure defects of the diaphragmatic muscle. This leads to protrusion of abdominal viscera into fetal chest and subsequent lung hypoplasia. This anatomical abnormality is usually suspected in routine second trimester ultrasound scans when a cystic structure (representing the stomach) is found in the thorax. In some cases mediastinal shift is the most prominent abnormality—this is more likely in right-sided diaphragmatic hernia. The diagnosis can be also easily done in the first trimester of pregnancy in cases of large defect. Right-sided hernias are more difficult to diagnose as the echogenicity of fetal liver may be similar to lung tissue. Prenatal diagnosis should prompt referral to a tertiary center for confirmation of the diagnosis, assessment of severity, and associated anatomic and genetic abnormalities. Invasive procedures, such as amniocentesis or cordocentesis are offered to confirm normal karyotype. In cases of isolated severe CDH, fetoscopic endotracheal occlusion (FETO) procedure may be beneficial and should be offered.[1-4]

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DIAGNOSIS

Most patients are referred to our center with suspicion of the defect. All patients are scanned and counseled by an experienced sonographer. The severity of CDH is established based on lung-to-head ratio (LHR) calculation.[5] Only in cases of suboptimal visualization or suspicion of coexisting anatomical abnormality, magnetic resonance imaging (MRI) is offered. All patients are also counseled by a geneticist and amniocentesis is performed to confirm normal karyotype. In all cases, cardiac scan is performed either by a fetal cardiologist or experienced sonographer. Patients with LHR <1 are considered as severe CDH and if isolated FETO procedure is offered. Before the procedure, observed-to-expected ratio is calculated, liver position established, fetal biometry and cervical length measured.

Fetoscopic Endotracheal Occlusion

Patients with LHR <1 who have decided to undergo FETO are admitted to the hospital before 27 weeks of pregnancy. After counseling and signing consent forms, the procedure is performed usually on the next day. Before the procedure, patients are advised to restrain from eating and drinking for at least 8 hours. They are administered with single dose of intravenous antibiotics (usually ampicillin) during or before the procedure. All interventions are performed in the surgical theater. The patient receives continuous epidural anesthesia. The position of the fetus and location of the placenta are

reevaluated by ultrasound. In some cases, fetus will be moved by external manipulation to obtain desired position. Fetal anesthesia is by either intramuscular injection or cordocentesis (pancuronium, fentanyl, atropine) with a 20 gauge needle under ultrasound guidance. A 12-french cannula will be inserted into the amniotic cavity under ultrasound guidance. The procedure will be performed with a miniature straight semirigid fetoscope 3.3 mm (11506 AAK, Storz). Detachable balloon (Golbal4, Balt, Extrusion) is placed in fetal trachea with the use of microcatheter (BALTACCIBDPE, Balt, Extrusion). The balloon is inflated with 0.7 ml of saline and detached 2 cm above the carina. The technical aspects of FETO procedure are similar as described by other authors.[6] After the procedure, patients are advised bed rest for at least a few hours. All patients are discharged from the hospital within the next few days and followed up every 2 weeks in ultrasound clinic until 32 weeks.

In cases of uncomplicated course (no preterm delivery, no premature rupture of membranes), patients are admitted to hospital at 32 to 33 weeks. Intramuscular corticosteroids (betamethasone) for lung maturation are administered (two injections 12 mg each) and on the next day balloon removal is planned. The procedure of balloon removal technically is similar to balloon occlusion, but the balloon is punctured (long needle) and removed from the trachea.

DELIVERY

In the most favorable course, delivery starts after 38 weeks. But the risk of premature rupture of membranes and/or preterm delivery is significant. Gestational age at birth is an important prognostic factor. After successful removal of the balloon, patients are transferred to multicenter hospital (tertiary reference) for the delivery and diaphragmatic repair.

DISCUSSION

Endoscopic tracheal occlusion for severe diaphragmatic hernia has been performed since 1998.[7] Over the last 20 years, many technical issues have been solved and improved, hence decreasing the risk of premature delivery and premature rupture of membranes. Fetoscopic endotracheal occlusion has been described as a potential procedure for increasing surviving rates for neonates with severe CDH. Once the diagnosis is made, multidisciplinary counseling about options, prognosis, and planning further management is necessary. In cases of severe and isolated CDH endotracheal balloon occlusion may be offered in centers with adequate equipment and trained specialists. So far in our center there have been 21 sets of successful tracheal occlusions and balloon removals. Considering our preliminary experience, the results are encouraging.

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