Bahrami Children's Hospital

**In the Name of God**

**Bahrami children Hospital**

دانشگاه علوم پزشکی و خدمات بهداشتی،درمانی تهران

| | | |
|---|---|---|
| **Date of notification:** 2020/SEP/17 | **congenital diaphragmatic hernia** | **Instruction code:** BH/INS/10 |
| **Edit number:1** | | **number of pages:2** |
| **Date Editor:2020/OCT/7** | | |

## What is congenital diaphragmatic hernia?

Congenital diaphragmatic hernia (CDH) occurs when there is a hole in the diaphragm, which is the thin sheet of muscle separating the chest from the abdomen. When this gap forms during a fetus's development in the womb, the bowel, stomach or even the liver can move into the chest cavity. The presence of these abdominal organs in the chest limits the space for the lungs and can result in respiratory complications. Because CDH forces the lungs to grow in a compressed state, several aspects of their function may not develop normally until after the birth of the baby.

## What causes CDH?

In a growing embryo, the diaphragm is completely formed by 10 weeks of gestation. However in cases of CDH, the process that leads to formation of the diaphragm is disrupted. Once there is a hole present in the diaphragm, abdominal contents can move into the chest. This is called **herniation**. Because fetal activity and breathing movements become more frequent and vigorous as a pregnancy continues, the amount of herniation can fluctuate or increase.

Sometimes CDH is caused by a problem with a baby's chromosomes or by a genetic disorder. If this is the case, the baby may have additional medical problems or organ abnormalities. In other instances, CDH may occur without an identifiable genetic cause. This is called **isolated CDH**, and under these circumstances the primary concern is the degree of pulmonary hypoplasia caused by the defect. In order to determine if CDH is isolated and to provide the most correct information about the disease, genetic testing is required.

## CDH Diagnosis

Detection of CDH may come during a routine ultrasound, which may reveal excess amniotic fluid and/or abdominal contents in the fetal chest cavity. To confirm a prenatal diagnosis of CDH, doctors may perform a very detailed ultrasound, conduct testing of the fetus's chromosomes and take measurements of its lung size. During the ultrasound examination, doctors focus on specific findings that may point to the presence of a syndrome. The genetic testing is performed by amniocentesis.

The lung size is then measured and compared to the expected size at this stage of a pregnancy. This can be done by measuring the lung area to head circumference ratio (LHR) or comparing the observed/expected LHR (o/e LHR). It is also important to determine

whether the liver has also moved into the chest. Based on these measurements, specialists at the Johns Hopkins Center for Fetal Therapy can grade the severity of CDH as mild, moderate or severe. Specialized imaging techniques including magnetic resonance imaging (MRI) are used to help achieve the most accurate assessment.

Congenital diaphragmatic hernia may also be diagnosed after birth — often if a newborn is having trouble breathing.

## CDH Treatment

Following delivery, a baby with CDH may undergo surgery to close the defect. However, surgery after delivery does not address the lung damage that has already occurred. For this reason, fetal therapeutic procedures are recommended in some pregnancies. These procedures may help decrease the amount of lung damage that can occur during the pregnancy. The goal of fetal treatment is to reverse some of the lung damage that results from compression of the lungs.

## Fetal Treatment for CDH

- **Fetoscopic tracheal occlusion (FETO)**: The fetal lungs produce fluid that leaves the body through the baby's mouth. If this outflow of fluid is blocked, it has nowhere to go and swells up in the affected lung. When this occurs over a period of four to five weeks, the lung expands and its function appears to improve. This type of blockage can be achieved by temporarily blocking the fetal windpipe (trachea) with a balloon for a period of time. This is done by performing operative fetoscopy, known as FETO. It is believed that FETO works by increasing the lung maturation and reversing some of the damaging effects of CDH on lung function.

- **Fetal surveillance and delivery planning**: There is a high possibility that a baby with CDH will get worse before the anticipated due date. Part of a comprehensive treatment plan will involve close fetal and maternal monitoring to avoid severe fetal deterioration and to determine the circumstances and timing for optimal delivery.

| Name / Signature Authors | Name / Signature Confirmers | Name / Signature Advising |
|---|---|---|
| **DR. Behzad Mohammadpour**<br>Assistant Professor<br><br>**Dr. Farid Eskandari**<br>Assistant Professor: surgery | **IPD team members** | **DR. Yousef Mojtahedi** |