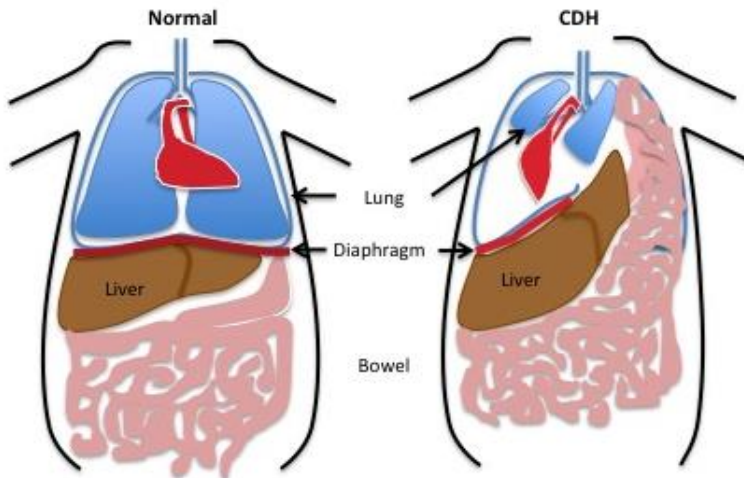


## Congenital Diaphragmatic Hernia: Important Things to Know

### What is congenital diaphragmatic hernia?

The diaphragm is a thin sheet of muscle that separates the chest from the abdomen. In the growing embryo, the diaphragm is completely formed by 10 weeks gestation. Approximately 1 in 4,000 fetuses have an inborn defect where the diaphragm is incompletely formed. The opening in the diaphragm may allow contents from the abdomen—including the bowel, stomach or even the liver—to enter the chest. This is called congenital diaphragmatic hernia or CDH. After birth, the hole in the diaphragm can usually be closed without difficulty. However, in fetal life, the abdominal contents that enter the chest



compress the lungs so they cannot develop normally in many ways. There are fewer tiny air sacs called alveoli that exchange oxygen and carbon dioxide. The distance between the air entering the lung and the blood vessels that take up the oxygen is increased. The number of blood vessels available to transport oxygen back to the heart is decreased. When the lungs are underdeveloped, it is called pulmonary hypoplasia. All of these factors decrease the ability for the lungs to function properly after birth.

The fetus does not need to use the lungs during pregnancy because

the placenta provides oxygen and nutrients. After delivery, many babies will need additional help to breathe. This can be provided by mechanical ventilation. Many forms of mechanical ventilation are available to support the baby after birth. However, if the pulmonary hypoplasia is too severe, the lungs may never be able to provide enough oxygen for the baby to survive. This is called lethal pulmonary hypoplasia. One of the major complications of CDH is the risk for lethal pulmonary hypoplasia.

Babies with CDH may also have other structural or genetic problems. A detailed ultrasound and genetic testing can help determine if this is your baby's only problem.

### What are the chances that my baby will survive?

The survival chances depend entirely on the development of the lungs prior to birth. Babies that have other structural or genetic problems can have additional challenges that lower chances for survival. Earlier and more severe compression of the lung tissue results in greater developmental problems. Therefore, survival can be predicted before birth by examining the lung size. The relative lung size can be assessed by two-dimensional ultrasound, three-dimensional ultrasound, or fetal magnetic resonance imaging (MRI). The ratio of the lung and the fetal head circumference (lung-to-head ratio = LHR) or the

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comparison of the observed to the expected size of the lung at each gestational age (observed/expected lung-to-head ratio = o/e LHR) are effective methods to predict the survival in CDH. Based on the LHR or o/e LHR and position of the liver, CDH can be divided into severe, moderate and mild categories. The survival rates are better in the mild and moderate categories, especially if managed at a center that is experienced in the care of babies with CDH before and after birth. At Johns Hopkins Hospital, the overall survival of babies with CDH is well above the national average of 68%. The assessment of severity before birth is also used to decide who may benefit from fetal treatment to improve chances of survival. All families that are planning to deliver a baby with CDH at Johns Hopkins Hospital meet with both the Neonatology and Pediatric Surgery teams during pregnancy.

### **Can my baby be treated?**

Under normal circumstances, the lungs produce fluid that can escape through the windpipe (trachea). Occlusion—or blocking—the trachea prevents escape of this fluid and stretches the lung tissue. This increase can stimulate lung growth. Extensive research has shown that fetal tracheal occlusion improves lung development in CDH. A technique has been developed where a small instrument with a miniature camera is used to look inside the uterus to insert a balloon into the trachea of the fetus. This procedure is called fetoscopic tracheal occlusion or FETO. This treatment is typically performed between 26-29 weeks gestation. A second procedure after 32 weeks is required to remove the balloon. After successful completion of these procedures, the baby can deliver vaginally. Because it is an invasive procedure, FETO is only performed for babies with severe CDH and no other structural or genetic problems.

### **How is FETO performed?**

This minimally invasive fetoscopic procedure is performed under epidural or local anesthesia. Prior to FETO, medication will be given to your baby for pain relief and also to prevent fetal movement. Then the fetus is gently moved into the best position. The fetoscope is introduced through the skin into the uterus and guided into the mouth and trachea of your fetus. When the fetoscope has been directed into the trachea, a small catheter is used to guide the balloon to its proper placement site. Once this is achieved, the balloon is inflated with fluid and left in the trachea. The balloon remains in the trachea of the fetus until 32-34 weeks gestation. During this time, your fetus will be monitored via ultrasound. Since the placenta provides the oxygen for the fetus, the balloon does not interfere with fetal breathing. Prior to birth, however, the balloon must be removed in order for the baby to be able to breathe. The balloon can be removed in several ways. While pregnant, a second fetoscopy may be performed. Alternatively, the balloon may be punctured under ultrasound guidance using a needle. Once the balloon is removed, you can deliver normally. If the baby is born before the balloon is removed, puncture can be performed after/at birth using a needle or by direct visualization of the balloon (tracheoscopy). Because of the special expertise needed, delivery in a tertiary care center with experience in the care of babies with CDH is important.

### **What are the risks for my baby and me?**

Since FETO is a minimally invasive procedure, the risks for the mother are small. Ultrasound is used to help avoid blood vessels and the placenta. The main risks are rupture of membranes or going into labor while the balloon is still in place. Under these circumstances, our dedicated FETO team performs emergent removal. Because this must be done quickly, patients receiving FETO must stay close to Johns



Hopkins Hospital while the balloon is in place. The balloon causes some bulging of the trachea, but this has not been shown to have any adverse effects and generally resolves after birth.

### **What are the alternatives?**

You may choose to have no intervention, or if your baby is not eligible for FETO, the pregnancy needs to be monitored closely with regular visits and ultrasound examinations. Your baby will need to be delivered at a tertiary care hospital experienced in the care of babies with CDH.

### **What happens after my baby is born?**

At birth, a breathing tube will be placed into the trachea to help with breathing and a small tube will be placed in the stomach to help the baby to avoid swallowing air. The baby will be admitted to the neonatal intensive care unit and placed on a ventilator to help the baby breathe as gently as possible. Over the first few

days, the baby is monitored to see how well she or he can exchange oxygen, release carbon dioxide and maintain blood pressure. The surgical repair of the hernia will be performed once the baby is stable or when determined by the multidisciplinary pediatric team. It is also important to know that babies with CDH may have additional difficulties with feeding and transition that require special attention during the newborn period and into childhood.

