Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia (CDH) is a developmental defect of the diaphragm. As the muscle that separates the organs in the abdomen from the organs in the chest, the diaphragm helps control breathing and is linked to the development of the lungs. CDH results from the abnormal development of the diaphragm before birth that causes a hole in the muscle. This condition affects about 1 in 5,000 babies. CDH may range from a small hole, or hernia, to the complete absence of the diaphragm.

An absent or partially formed diaphragm allows the stomach and intestines to move into the chest cavity during pregnancy and crowd the baby’s heart and lungs. This can lead to compression and underdevelopment of the lungs (pulmonary hypoplasia), and potentially life-threatening breathing difficulties after birth. CDH may also cause high blood pressure in the arteries of the lungs (pulmonary hypertension), requiring your baby’s heart to work harder to pump blood to the arteries. Over time, the heart may weaken, resulting in heart failure.

Diagnosis and Prenatal Monitoring

Diagnosis of CDH usually occurs in the second trimester, when a routine prenatal ultrasound shows that abdominal organs have moved into the chest area. When CDH is suspected, patients are often referred to a maternal-fetal specialist for further care and evaluation. Maternal-fetal medicine physicians affiliated with The Fetal Center at Children’s Memorial Hermann Hospital may recommend other tests, including fetal MRI and fetal echocardiogram to assess the severity of the condition. They may also recommend amniocentesis to identify possible chromosomal abnormalities.

During pregnancy, maternal-fetal medicine specialists monitor mother and baby to ensure that the baby develops appropriately in the uterus. The team of specialists at The Fetal Center work closely with families to determine the best course of treatment before and after delivery. This includes affiliated pediatric surgeons and affiliated neonatologists, as well as genetic counselors. A coordinator keeps mothers in contact with all of the appropriate physicians and specialists and coordinates all aspects of care. Families are educated on CDH as well as its treatment options and possible outcomes.
Treatment After Delivery
Patients and families with a CDH fetus will undergo a comprehensive birth plan developed by The Fetal Center team. The timing and type of delivery is determined by the severity of the CDH through prenatal evaluation. This could include delivery around 37 weeks gestational age by either cesarean section or vaginal delivery. Careful planning allows the CDH team to be prepared for the birth of the baby.

Babies with CDH can develop a wide range of disease severity. Some infants have normal lung function and often do not require much assistance. Others can suffer from extreme respiratory failure with dismal outcomes. The team at The Fetal Center and Children’s Memorial Hermann Hospital considers every patient a potential survivor. As such, the team undertakes a very aggressive approach in the pre- and postnatal management of CDH, operating on 93 percent of all patients. As a result, outcomes are in the top 10 percent worldwide, with lower-than-expected pulmonary morbidity.

When newborns with CDH suffer from the most severe cases, they may require external oxygen support via a form of heart-lung bypass, known as extracorporeal membrane oxygenation (ECMO). The ECMO program at Children’s Memorial Hermann Hospital was the first in Houston and one of the first in the nation to provide the therapy for critically ill babies with respiratory failure. The Extracorporeal Life Support Organization has named the program a “Designated Center of Excellence” continuously since 2006.

Once the baby’s breathing has been stabilized, which may take days or weeks, surgical repair of the hole in the diaphragm is performed. Surgery returns the abdominal organs to the abdominal cavity and makes room in the chest for the compressed lung to grow.

Based on the baby’s condition and the severity of CDH, pediatric surgeons may use a minimally invasive approach with small incisions to perform the repair. In mild cases of CDH, the surgeon may repair the hernia by just bringing the diaphragm muscle together. In more severe cases, the defect may be very large; often the defect is repaired using surgical mesh or patch, which may need to be replaced as the child grows.

Long-term Follow-up at the CDH Clinic
Complications and long-term outcomes depend greatly on the severity of the defect. Following delivery, CDH repair and discharge from the hospital, a team of pediatric specialists continue to care for CDH babies. Some babies will mature normally without lifelong complications; however, most babies experience some long-term respiratory, gastrointestinal, neurological or other health issues that require close follow-up and timely treatment.

Because CDH is a developmental defect with many facets, newborns and children with the disorder are followed in the Congenital Diaphragmatic Hernia Clinic, where they have access to specialized multidisciplinary care.

The Congenital Diaphragmatic Hernia Registry
McGovern Medical School at UTHealth is home to the Congenital Diaphragmatic Hernia Study Group and the international CDH Registry, a voluntary collaborative database that has gathered data on more than 9,000 babies with CDH since it was founded in 1995. The CDH study group was founded and led by Dr. Kevin P. Lally, Chair of Pediatric Surgery at McGovern Medical School and Surgeon-in-Chief at Children’s Memorial Hermann Hospital. The registry represents children’s hospitals in 14 countries, and its presence ensures that physicians provide the most up-to-date data for CDH outcomes.

To refer a patient for evaluation for FETO as part of the TOTAL trial, call 832.325.7288.
## Meet the CDH Team

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