A GUIDE TO SPECIALIZED CARE OF CONGENITAL DIAPHRAGMATIC HERNIA

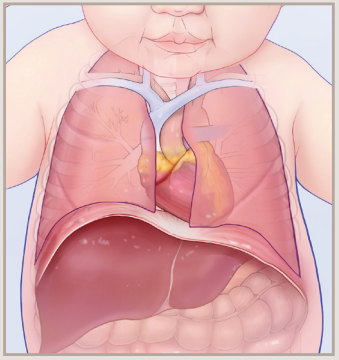
Children's Hospital of Philadelphia®
Richard D. Wood Jr.
Center for Fetal Diagnosis & Treatment

SPECIALIZED CARE OF CDH

Finding out that an unborn baby has a birth defect such as congenital diaphragmatic hernia (CDH) can be overwhelming. Learning about CDH is the first step to understanding what is going on with the baby and what care might entail. This educational guide was developed by our specialized team at Children's Hospital of Philadelphia's Richard D. Wood Jr. Center for Fetal Diagnosis and Treatment (CFDT) to help parents understand this condition and begin to orchestrate the best care for themselves and their baby.

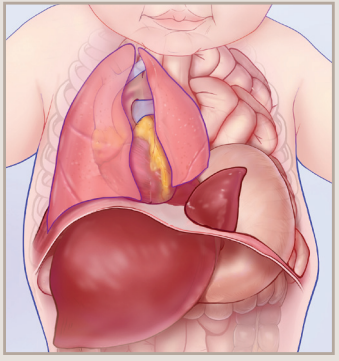
FACTS ABOUT CDH

- About 1,600 babies are born with CDH every year in the United States, or 1 in every 2,500 live births. The same number of babies are born with cystic fibrosis or spina bifida.
- The diaphragm muscle separates the heart and lungs in the chest from the abdomen. CDH occurs when a hole in the diaphragm muscle fails to close and the contents from the abdomen (stomach, intestine and/or liver) migrate into the chest through this hole.
- When the abdominal organs are in the chest, there is limited room for the lungs to grow, resulting in pulmonary hypoplasia (or underdeveloped lungs).
- CDH can occur on the left side, right side or, very rarely, on both sides.



NORMAL DIAPHRAGM ANATOMY

The diaphragm is a muscle that separates the heart and lungs from the abdominal cavity and supports breathing.



CDH ANATOMY

In CDH, a hole in the diaphragm allows abdominal organs to move into the chest, restricting lung development.

EVALUATION AT THE CENTER FOR FETAL DIAGNOSIS AND TREATMENT

A COMPREHENSIVE MULTIDISCIPLINARY EVALUATION

After a doctor has diagnosed CDH in a pregnancy, the next step is to be referred to a prenatal diagnosis center for additional testing and information. If a patient is referred to our CFDT, a comprehensive multidisciplinary evaluation will be scheduled, and all of the following tests would occur in one day.

- **High-resolution level II ultrasound** is performed by our sonographers and fetal radiologists and often lasts up to two hours. The ultrasound will confirm the diagnosis and the side of the hernia, identify any additional malformations, determine lung size, and calculate a lung-to-head ratio (LHR) and an observed over expected ratio (O/E) for LHR. The ultrasound also determines the position of the liver.

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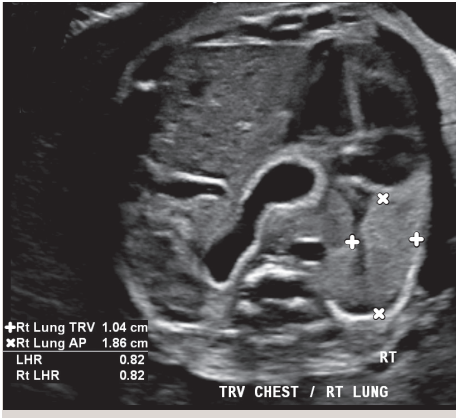

High-resolution ultrasound of a congenital diaphragmatic hernia.

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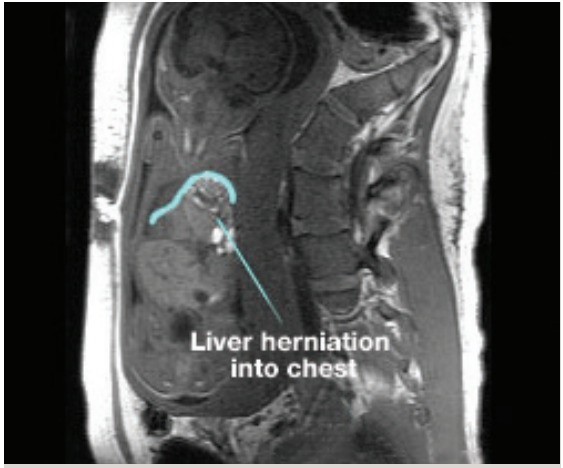
EVALUATION AT THE CENTER FOR FETAL DIAGNOSIS AND TREATMENT

- **Fetal echocardiogram** or targeted ultrasound of the baby's heart is performed by our fetal cardiologist. The fetal echo determines if there are any structural defects of the heart. It can sometimes be difficult to perform when the abdominal organs are in the chest causing the heart to be compressed.
- **Ultrafast fetal MRI,** a technique pioneered at Children's Hospital of Philadelphia (CHOP), provides further anatomy detail regarding the liver position and total lung volume. Large amounts of liver in the chest decrease O/E lung volumes.



This ultrafast fetal MRI confirms liver herniation into the chest.

PATIENT COUNSELING & EDUCATION

Patients will speak with one of our nurse coordinators before their first appointment. This same coordinator will review diagnosis-related education, answer any questions the patient may have and discuss the plan for their one-day evaluation.

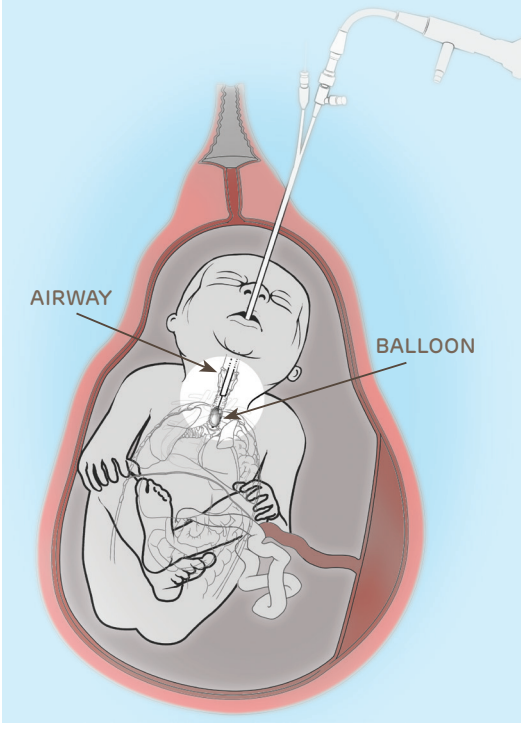
The patient will also meet with a genetic counselor, who will review the family history and prenatal genetic testing.

Once the diagnostic exams are complete, the patient will meet with a high-risk maternal-fetal medicine specialist (MFM), a pediatric surgeon and the coordinator. They will review the maternal health history and the results of all exams. In addition, they will discuss all treatment options and prenatal and postnatal care, as well as delivery recommendations.

FOR SEVERE CDH

FETOSCOPIC ENDOLUMINAL TRACHEAL OCCLUSION (FETO)

- Through a small incision in the mother's abdomen, a balloon will be placed in the unborn baby's airway between 29 to 30 weeks gestation.
- Blocking the airway causes fluid to build up in the unborn baby's lungs; pressure from the fluid, in turn, causes the lungs to grow. Bigger lungs may improve survival after birth.
- A second procedure occurs around 34 weeks to remove the balloon so the baby can breathe at birth.
- While the balloon is in place, the airway is blocked. As a result, if an unexpected delivery occurs, special management of the airway is required. For this reason, mothers enrolled in this study must remain near our fetal center from the time of balloon placement through delivery in our Special Delivery Unit and the baby's discharge from our Newborn/Infant Intensive Care Unit.
- Children who undergo FETO still require surgery to repair the CDH after birth, and their postnatal care and follow-up is similar to children who do not undergo the fetal procedure.



PRENATAL MANAGEMENT

Follow-up ultrasound exams should be performed every four weeks throughout the pregnancy to evaluate the baby's growth and activity. If a patient lives at a distance from Philadelphia, these ultrasounds can be performed locally. At 34 weeks, ultrasounds increase to weekly.

Relocation (to the Ronald McDonald House, a Hosts for Hospitals site or another partner site) is necessary if a patient lives more than one hour away from our center at about 34 weeks or sooner if extra fluid develops around the baby (polyhydramnios) or there are signs of preterm labor. This will ensure the patient is nearby in the event labor occurs earlier than expected.

OUR TEAM WILL MONITOR THE PREGNANCY FREQUENTLY.*

**Monitoring closely will help us prevent poor outcomes.*

DELIVERY

If mom and baby meet the qualifications, a vaginal delivery at term is planned in our Special Delivery Unit (SDU), which was created specifically for the healthy mother whose pregnancy is affected by a birth defect. The majority of babies with CDH can be delivered vaginally. However, when a woman is carrying a baby with CDH, the threshold for making the decision to deliver by C-section is lower (C-section is more likely). If a woman has any maternal health issues, delivery at the adjacent Hospital of the University of Pennsylvania will be arranged.

The first hour after a baby with CDH is born is an especially critical transition period. It is referred to as "the golden hour." A baby must go from being completely supported by its mother, into the world, where the baby has to breathe and survive all on its own. Most babies with CDH cannot make this transition from womb to world on their own. The SDU has an expert team dedicated to helping babies during this precious hour. We deliver the largest number of critically ill infants with birth defects in the nation.

After your baby is born, they are carried to our stabilization suite, where our expert Newborn Stabilization and Resuscitation Team provides all the support your baby needs before they are taken to the N/IICU for further care.

GOLDEN HOUR

The Newborn Stabilization and Resuscitation Team plays a critical role in our delivery room care. This team is made up of a dedicated group of clinicians who are skilled in the delivery room care of high-risk babies with known congenital anomalies (both cardiac and non-cardiac). The team works closely with maternal-fetal medicine (MFM) specialists and obstetricians in the Special Delivery Unit to prepare for a baby's arrival. They use advanced monitoring to evaluate every aspect of a baby's health from the moment they are born.

The team's goal is to optimize care of the newborn immediately after birth to improve outcomes. They have developed evidence-based tools to guide golden hour care for each type of diagnosis, including CDH. They also created an interactive critical knowledge platform that integrates with patient monitors and equipment.

SPECIALIZED NEONATAL SURGICAL TEAM

- It is important that the baby be treated by a team with experience caring for babies with CDH. At CHOP, we see more babies with CDH than anyone in the U.S.

- The baby will be cared for in the N/IICU using optimal care guidelines developed by our experienced multidisciplinary team.

- The baby may or may not require specialized equipment such as the oscillator ventilator, heart lung machine (ECMO) or nitric oxide, but it is important that they have immediate access when necessary.

OUR NEONATAL SURGICAL TEAM IS THE ONLY TEAM OF ITS KIND IN THE WORLD, MADE UP OF SPECIALISTS WHO HAVE WORKED TOGETHER SINCE 2004.

SURGERY FOR CDH

- Babies with CDH are extremely sensitive to noise and movement, so surgery is often performed in the N/IICU so the baby does not have to be transported to the operating room.
- The baby will receive general anesthesia and will be continually monitored by a pediatric anesthesiologist.
- An incision is made just below the baby's rib cage, the organs in the chest are guided back down into the abdomen and the hole in the diaphragm is sewn closed. The space created in the chest allows the lungs to continue to grow; children will continue to grow more air sacs, or alveoli, through early childhood.

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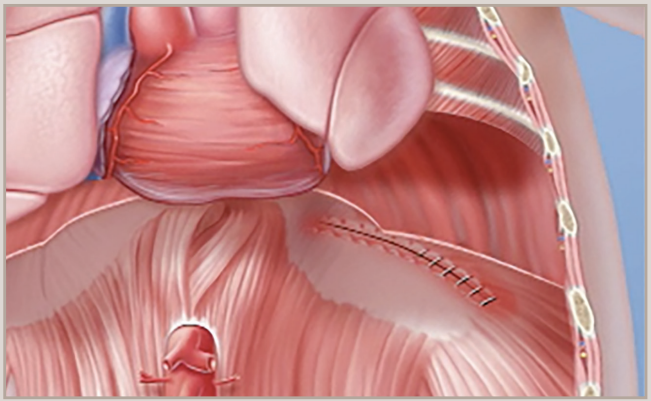

To repair the hole in the diaphragm, an incision is made just below the baby's rib cage and the organs in the chest are guided back down into the abdomen.

The muscles around the hole are then sewn together. The space created in the chest allows the lungs to continue to grow.

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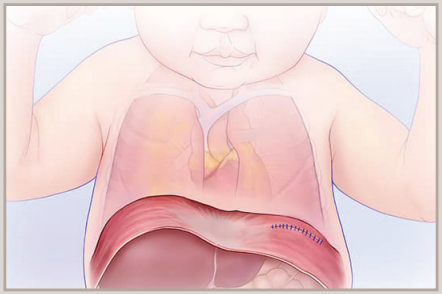
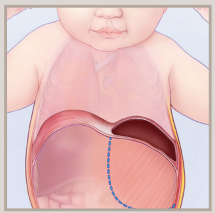
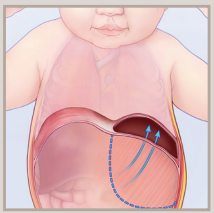
SURGERY FOR CDH

- For those babies with large defects or completely lacking a diaphragm, the hole is closed with either a muscle flap or a GORE-TEX® patch.
- Sometimes the abdominal wall cannot be closed during surgery. In these cases, temporary placement of a silo or mesh may be recommended.

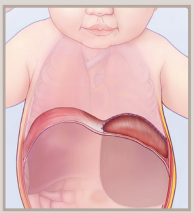


After gently bringing the liver, stomach and intestines into place, a surgeon sews the muscles around the hole together. The space created in the chest allows the lungs to continue to grow.





For babies with a larger hole in the diaphragm, a muscle flap is used to cover the hole.



The large hole is sewn closed by attaching the muscle flap to the diaphragm muscle.

ECMO

Babies with CDH may require extracorporeal membrane oxygenation (ECMO).

- Under sterile conditions and once a baby has received pain medication, the pediatric surgeon will place two tubes called cannulas into the artery and vein in the baby's neck at the bedside. The tube in the neck drains blood out of the body from the large vein and sends it to the ECMO circuit where it is oxygenated. The other tube returns the now-oxygenated blood to the baby by the carotid artery.

- ECMO is used when other treatments are unsuccessful. The lungs rest as the ECMO circuit does the work.

- In some cases the baby may have the CDH repair while on ECMO.

- ECMO can have serious complications, including bleeding and infection. Careful monitoring by an experienced ECMO team is critical.

- One ECMO specialist and an experienced neonatal surgical registered nurse oversee care at all times.

- CHOP has been designated a Platinum Center of Excellence by the Extracorporeal Life Support Organization since 2008.

- Our program has done more than 1,700 ECMO treatments since beginning in 1990, with the team providing ECMO support to over 300 CDH runs.

LONG-TERM FOLLOW-UP

Long-term follow-up by a team of experts is important to provide the best clinical care, and also to improve the understanding of pulmonary hypoplasia so that we can continuously improve counseling and care.

- As of 2023, the Pulmonary Hypoplasia Program (PHP) at Children's Hospital of Philadelphia follows more than 1,200 children, approximately 65% of whom were born with CDH, well into school age.
- The team includes clinicians from general surgery, developmental pediatrics, pulmonary, cardiology, psychology, nutrition, audiology, social services and others as needed.
- Initial follow-up by the PHP will be arranged when the baby is in the N/IICU.
- Appointments are tailored to each child's needs, but typically occur at 6 months, 12 months, 2 years, 4.5 years and 6 years, and then every two years thereafter, as needed. Appointments are designed for visits to multiple specialists in one day.
- CHOP is committed to long-term partnership by evaluating neurodevelopmental outcomes in children born with CDH so that we can continuously improve care and better understand long-term outcomes.

OUR PULMONARY HYPOPLASIA PROGRAM IS A UNIQUE PROGRAM THAT PROVIDES COMPREHENSIVE, INTERDISCIPLINARY FOLLOW-UP CARE FOR CHILDREN WITH CDH.

KEY RESOURCES

CHILDREN'S HOSPITAL OF PHILADELPHIA

Richard D. Wood Jr. Center for Fetal Diagnosis and Treatment
fetalsurgery.chop.edu/cdh
1-800-IN UTERO (468-8376)

PULMONARY HYPOPLASIA PROGRAM

chop.edu/php
215-590-2733

CDH INTERNATIONAL (CDHi)

cdhi.org

PEYTON'S PROMISE

peytonspromise.com

Our team has seen an exceptionally high volume of babies with CDH since 1995 and we are among the most experienced programs in the world in caring for pregnancies complicated by this condition.

SCAN THIS CODE TO LEARN MORE ABOUT OUR VOLUMES AND OUTCOMES.

1-800-IN UTERO (468-8376)
or 215-590-5190

fetalsurgery.chop.edu

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