



# A GUIDE TO SPECIALIZED CARE OF CONGENITAL DIAPHRAGMATIC HERNIA



**Children's Hospital  
of Philadelphia®**

Center for Fetal Diagnosis & Treatment

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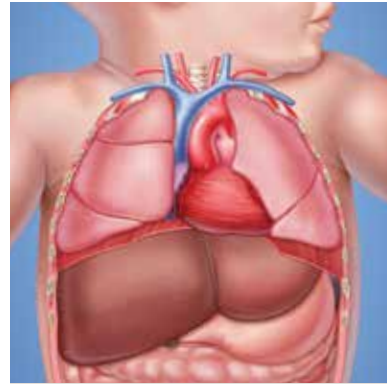
# SPECIALIZED CARE OF CDH

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 Center for Fetal Diagnosis and Treatment to help parents understand this condition and begin to orchestrate the best care for themselves and their baby.



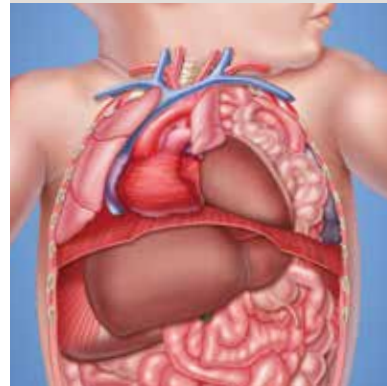
# FACTS ABOUT CDH

- 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

The diaphragm is a sheet of muscle that separates the heart and lungs from the abdominal cavity.



## With CDH

In CDH, a hole in the diaphragm allows abdominal organs to move into the chest and restrict 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 Center for Fetal Diagnosis and Treatment (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, which is very important.

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High-resolution ultrasound of a congenital diaphragmatic hernia.

# EVALUATION AT THE CENTER FOR FETAL DIAGNOSIS AND TREATMENT

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- **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

Upon arrival to our center, the patient will be greeted by one of the nurse coordinators or genetic counselors she initially spoke with when she contacted us. The coordinator will review any questions the patient may have, discuss the schedule for the day, and provide a tour of the Harriet and Ronald Lassin Newborn/Infant Intensive Care Unit (N/IICU) and the Garbose Family Special Delivery Unit (SDU), as requested.

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 (MFHM), a pediatric surgeon and her 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.



# FETOSCOPIC ENDOLUMINAL TRACHEAL OCCLUSION (FETO)

Severe CDH can result in serious disease and death. To help improve outcomes in patients with severe pulmonary hypoplasia — defined as O/E LHR < 25% — our team is part of a research study of a fetal treatment that may allow the lungs to grow enough before birth so that these children are capable of surviving and thriving.

- A minimally invasive procedure for severe CDH cases.
- Eligible candidates may undergo two prenatal procedures.
- Gestational age < 30 weeks.
- The first procedure occurs between 27<sup>0</sup> and 29<sup>6</sup> weeks gestation.
- Through a small incision in the mother's abdomen, a balloon will be placed in the unborn baby's airway.

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# FETOSCOPIC ENDOLUMINAL TRACHEAL OCCLUSION (FETO)

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- The balloon is inflated until it blocks the airway and is left in place for a few weeks.
- 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.
- The second procedure occurs around 34<sup>0</sup> to 34<sup>6</sup> 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 not enrolled in the study.



# 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 one of our Ronald McDonald Houses 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.



IN CONSULTATION WITH A PATIENT'S DOCTOR, OUR TEAM WILL MONITOR THE PREGNANCY FREQUENTLY, ESPECIALLY IN THE LAST TRIMESTER.\*

*\*Monitoring closely will help us deliver earlier, if necessary.*

# DELIVERY

The majority of babies with CDH can be delivered vaginally. However, there is a low threshold for performing a C-section delivery for fetal indications. If a woman has any maternal health issues, delivery at the adjacent Hospital of the University of Pennsylvania will be arranged.

If mom and baby meet the qualifications, a vaginal delivery at term is planned in our SDU, which was created specifically for the healthy mother whose pregnancy is affected by a birth defect. At delivery, the baby is carried to a pass-through window into a room where the Neonatal Surgical Team — including neonatologists, surgical advanced practice nurses, neonatal and surgical fellows, neonatal surgical nurses, and respiratory therapists — is waiting to stabilize the baby before bringing them to the mother and then to the N/IICU for further care.

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# DELIVERY

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At stabilization, a breathing tube is inserted into the baby's airway so a ventilator can be attached to help with breathing. A tube going from the mouth or nose to the stomach will be placed to keep air from getting into the stomach and intestines causing less space for the lungs to expand. Intravenous and arterial lines will be placed in blood vessels of an extremity or the belly button for administration of fluids and medications. Blood gases will be drawn from the arterial line to assess how well the baby can oxygenate.



# 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 nearly 50 babies with CDH every year – more than anyone in the U.S.
- The baby will be cared for 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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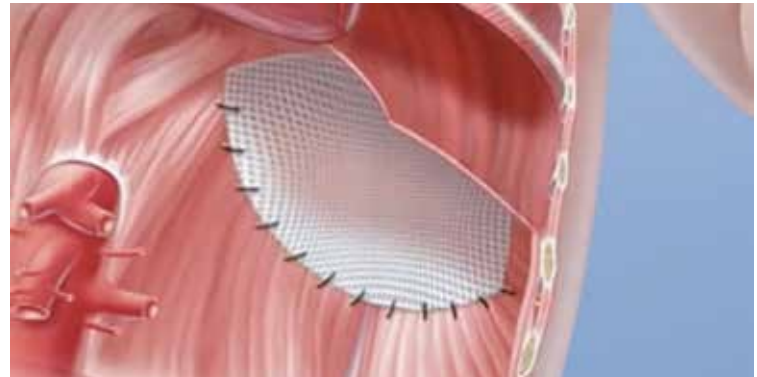
To repair the hole in the diaphragm, an incision is made just below the baby's rib cage and the organs in the chest are guided back down into the abdomen.

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

# SURGERY FOR CDH

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- For those babies with large defects or completely lacking a diaphragm, the hole usually is closed with 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.



For babies with larger defects, the hole is sewn closed with a soft tissue patch. The condition of the patch is monitored through long-term follow-up.

# ECMO

Babies with CDH who have severe pulmonary hypoplasia or fragile lungs 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 specialist is critical.
- One ECMO specialist and an experienced neonatal surgical registered nurse oversee care at all times.
- CHOP has been designated a Center of Excellence by the Extracorporeal Life Support Organization since 2008.
- Our program has done more than 1,440 ECMO treatments since beginning in 1991, with the team providing ECMO support to more than 245 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.

- The Pulmonary Hypoplasia Program (PHP) at Children's Hospital of Philadelphia follows more than 800 children, most of whom were born with CDH, well into school age.
- The team is made up of 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 for these children.

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



# CHOP'S CDH EXPERIENCE

From all over the United States and  
around the world since 1995.

Patient referrals

OVER  
**1,560**

Surgical repairs

OVER  
**500**

SDU deliveries

OVER  
**350**

ECMO runs

OVER  
**245**

For the most up-to-date information  
on our volumes and experience, visit  
[fetalsurgery.chop.edu/cdh](http://fetalsurgery.chop.edu/cdh)



OUR TEAM HAS SEEN AN  
EXCEPTIONALLY HIGH VOLUME  
OF CDH BABIES SINCE 1995 AND  
PROVIDES AN UNPARALLELED LEVEL  
OF CONTINUOUS EXPERIENCE.

*Center for Fetal Diagnosis & Treatment at CHOP, data on file as of 6/2019*

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# KEY RESOURCES

## CHILDREN'S HOSPITAL OF PHILADELPHIA

Center for Fetal Diagnosis and Treatment

[fetalsurgery.chop.edu/cdh](http://fetalsurgery.chop.edu/cdh)

1-800-IN UTERO (468-8376)

## PULMONARY HYPOPLASIA PROGRAM

[chop.edu/php](http://chop.edu/php)

215-590-2733

## CDH INTERNATIONAL (CDHi)

[cdhi.org](http://cdhi.org)

## BREATH OF HOPE

[breathofhopeinc.com](http://breathofhopeinc.com)

## PEYTON'S PROMISE

[peytonspromise.com](http://peytonspromise.com)

# CONTACT US

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

[fetalsurgery.chop.edu](http://fetalsurgery.chop.edu)



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