

Congenital Diaphragmatic Hernia

What is Congenital Diaphragmatic Hernia (CDH)?

Congenital diaphragmatic hernia is a fetal defect that develops during the formation of the diaphragm; this is the muscle that separates the chest cavity from the abdominal cavity. The diaphragm is the muscle that helps us to breathe, so it is a critical component of the respiratory system. In a diaphragmatic hernia, the diaphragm which develops at about 8 to 10 weeks of pregnancy does not completely form, and an opening remains between the chest and the abdominal cavities.

The severity of this condition ranges from a small opening in the diaphragm to no diaphragm on one side of their body. Ninety percent of all defects are found on the left side of the diaphragm. Depending on how large the hole is the intestines, spleen, liver and/or stomach may move up into the chest cavity. In turn, this causes the lungs to develop abnormally.

A CDH is a life-threatening abnormality. It is estimated that 1 in every 2,200 births is affected by diaphragmatic hernia. Parents that have one child born with a diaphragmatic hernia are at an increased risk of having another child with this condition; their chance is approximately 1 in every 50. The exact cause is not known at this time. Approximately 25% of babies with CDH will have another birth defect; most commonly cardiac and chromosomal. The survival rate with no other associated anomalies is about 80%.



Dr Monja Proctor



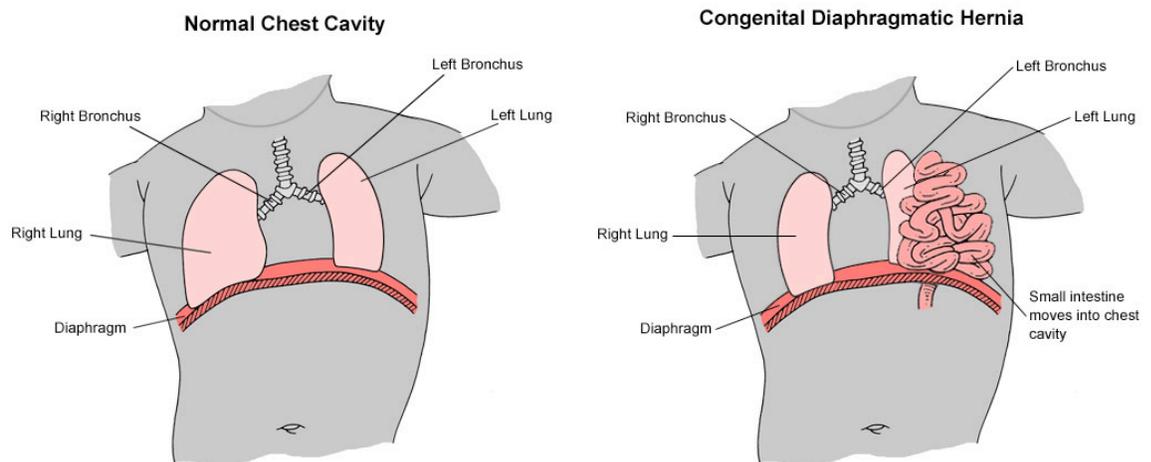
Dr Edwin Hatch



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How is a congenital diaphragmatic hernia diagnosed?

In many cases CDH is diagnosed during a prenatal ultrasound. If not diagnosed prenatally, the condition may be found soon after birth when the baby has trouble breathing. Less frequently, a diaphragmatic hernia is found in an older child.

How is CDH treated?

Surgery is needed to move the organs from the chest back down into the abdominal cavity and to close the hole in the diaphragm. Surgery is usually done within the first two weeks of life. Your baby will be cared for in Neonatal Intensive Care Unit (NICU) right after delivery. Most infants will be on a breathing machine called a *ventilator* before and after surgery to help with breathing. Babies with diaphragmatic hernia usually have small, stiff lungs. Over time your child's lungs will grow and the stiffness will improve. Oxygen and medications are used to help in the beginning. Surgery is delayed until the lung function is optimized as surgery in the short term can make breathing more difficult. Rarely infants require ECMO (Extra Corporeal Membrane Oxygenation) which is a type of bypass machine that allows blood to receive oxygen outside the lungs. If your child needs ECMO then your child will be transferred to another facility.

Will my baby be in pain?

The hole in the diaphragm and position of the intestine in the chest at birth does not hurt. This is simply the way the organs develop. The surgical team will ensure your baby is pain free before and after surgery with the aid of pain medication.

How serious is CDH?

It depends upon the size and development of the lungs. If the hole in the diaphragm is small and the lungs are of good size and function, your child may have little trouble breathing after surgery. If the hole is larger and the lungs are small and stiff, your child will require prolonged breathing support and a longer hospital stay.

Preparing before the baby's birth

You can begin preparing for the birth of your baby now through prenatal counseling with the Pediatric Surgery team at Swedish. This provides an opportunity to know the team who will be caring for your baby. A tour of the hospital and NICU can be arranged. Parent support groups like CHERUBS are available. You may also want to create a birth plan with your obstetric team who will deliver your baby.

What happens when my baby is born?

Immediately after delivery your baby will be examined by specialized pediatricians called Neonatologists. Your baby will be transferred to the neonatal intensive care unit (NICU). If your baby is having difficulty in breathing a ventilator will be used to help your baby breathe. Your baby may need a ventilator until after surgery. An intravenous line (IV) is placed to give fluids, nutrition and medicines directly into a vein. A nasogastric (NG) tube is placed in the stomach through the nose to empty the stomach and prevent distention. Your baby's blood pressure, breathing and oxygen levels will be continuously monitored.

What about feeding my baby?

If you are planning to breastfeed your baby, the staff will do everything they can to support your breast feeding goals. It is important to start breast pumping soon after your baby is born. Lactation nurses will be available to help.

During the evaluation period in the NICU and the first few days after surgery, it may not be safe to feed your baby. Feedings by mouth require a lot of energy and coordination. This can be stressful for babies with breathing problems from a diaphragmatic hernia. All your baby's nutrition needs will be provided through an IV in the form of TPN (Total Parenteral Nutrition). Once your baby is ready for feeding, either formula or your breast milk will be slowly introduced. Gradually as your baby tolerates the feeds your baby will be given more feeds and less IV nutrition (TPN).

Once your baby can handle a reasonable amount at a time you may be allowed to start breastfeeding. Nurses and infant feeding therapists will help with the feedings.

How long is the hospital stay?

Every child's recovery is different. Your baby will be ready to go home once he/she is breathing comfortably and is getting sufficient nutrition and is gaining weight. Most newborns with CDH are in the hospital for several weeks to several months depending on the recovery. Some babies may go home on oxygen to support breathing, or with a feeding tube called a gastrostomy tube to help with feedings.

What is the long term outlook for my baby?

Some children with this condition may have some long term issues around feeding or breathing, or developmental delay. Your child will be followed closely to manage any long-term issues.

Where can I get more information?

Contact CHERUBS, a parent support group offered through The Association of Congenital Diaphragmatic Hernia Research Advocacy and Support

CHERUBS
P.O Box 1150
Creedmoor, NC 27522
www.cherubs-cdh.org

Please call the pediatric surgery office at 206-215-2700 if you have any questions.

Notes



Pediatric General Surgery
1101 Madison, Suite 800
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For a free physician referral:
1-800-SWEDISH (1-800-793-3474)
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