

John B. Pietsch, M.D. William F. Walsh, M.D. Kelly Bennett, M.D.

DIAPHRAGMATIC HERNIA

What is a congenital diaphragmatic hernia (CDH)?

The diaphragm is a large dome-shaped muscle that separates the chest from the abdomen. The diaphragm normally develops between the seventh and tenth weeks of pregnancy.

An abnormal opening or hole in the diaphragm is called a diaphragmatic hernia. This opening allows some of the baby's abdominal organs to move into the chest. The stomach, small intestine, spleen, part of the liver, and the kidney can enter the chest cavity. The intestine takes up some of the space where the lung should develop, so that the lung can not develop normally. When this happens, we say that the lung is "hypoplastic." The more space the abdominal organs take up in the chest, the more severe the effects on the lung.

What causes a diaphragmatic hernia?

The abnormal opening ("hernia") is caused when the diaphragm muscles do not grow together normally during the baby's development. We do not yet know why this occurs, or how to prevent this from happening. The two types of diaphragmatic hernia are based on which side of the chest is affected.

- **Left sided hernias** are called Bochdalek hernias. 90% of hernias are on this side and they are slightly more common in boys. This type of hernia can be due to the diaphragm not forming properly or the intestine becoming trapped in the chest cavity as the diaphragm forms.
- **Right sided hernias** are known as Morgagni hernias. 2% of hernias are on this side and they are more common in girls. This type of hernia

is caused when the tendon in the middle of the diaphragm does not develop properly.

Could my baby have any other abnormalities?

About 1 in 3 infants with diaphragmatic hernia will also have another birth defect. In these cases, it is most often a heart defect. Rarely the baby may have a chromosomal abnormality such as trisomy 21 or Down syndrome.

What are the chances this will happen with my next baby?

There is a 2% chance of your having another baby with congenital diaphragmatic hernia (CDH). This means there is a 98% chance that the condition *would not be* seen in a future pregnancy.

How is CDH detected?

There are usually no signs or symptoms in the pregnancy. The pregnant mother may have an increased amount of amniotic fluid surrounding the baby. This excess of fluid is called “polyhydramnios.” The fetal ultrasound may show abdominal organs in the chest cavity, but the defect is not always seen on routine ultrasound. Because of this, some infants are not diagnosed until after they are born.

How will this condition affect my baby’s health?

A diaphragmatic hernia is a life-threatening condition. The diaphragm muscle is not properly formed and allows the lung to be crowded by other organs. Because of this crowding, these babies have serious difficulty breathing. Your baby will need efforts to help his or her breathing and heart function immediately after birth and during and after surgery. Even with excellent care, about 25% of babies with diaphragmatic hernia live for only a short time.

Treatment during pregnancy:

On rare occasions would a baby with CDH be considered eligible for surgery before birth. Any surgery before birth is called “fetal intervention.” Whether your baby is eligible or not depends on three things:

1. The side of the chest most affected by the diaphragmatic hernia
2. The time of diagnosis
3. The severity of the problem for the lung.

How will CDH affect my baby’s birth?

Many babies born with this condition will experience common signs, but babies may have different symptoms. Babies often are unable to get enough air or the oxygen it contains shortly after birth. This is called “respiratory distress,” and occurs because the diaphragm does not move properly and because the lung is crowded by the organs. The baby’s doctor will examine him or her for the following signs:

- abnormal chest development with one side larger than the other
- breath sounds absent on the affected side
- bowel sounds heard in the chest
- abdomen that appears to be caved in (concave) and feels less full to the touch
- severe breathing difficulty
- bluish coloration of the skin due to lack of oxygen
- fast breathing (tachypnea)
- fast heart rate (tachycardia)

If the baby is in enough distress they will be “intubated” with a breathing tube in order to help them breathe and a “replogle” will be placed to get rid of extra air in the stomach.

What will happen after my baby is born?

Several studies may need to be performed to evaluate the baby:

- Chest x-ray may show abdominal organs in the chest cavity

- A blood test will be performed to evaluate the baby's breathing ability
- Echocardiogram (to evaluate the heart)

Will my baby need surgery later?

Babies born with this condition will need treatment in a neonatal intensive care unit (NICU). After birth, surgery is needed to place the abdominal organs into the abdominal cavity and to repair the opening in the diaphragm. Most centers wait until the baby is stable before doing surgery. Some babies are good candidates for placement on a modified heart/lung bypass machine called ECMO (extracorporeal membrane oxygenation). The medical team and family together discuss whether this machine will improve the baby's outcome. ECMO gives the lungs more time to recover and function normally. Babies who require ECMO due to the severity of CDH have a 50% survival rate.

Expected Progress:

The infant's lung tissue on the affected side may be underdeveloped, and the infant's progress depends upon the development of the lung tissue. Generally, the prognosis is good for infants who have adequate lung tissue. With advances in neonatal and surgical care, overall survival is now greater than 80%. The infant may require several weeks of hospitalization after surgery, depending on how long breathing needs to be supported by a ventilator. Feeding begins after the first bowel movement is passed. Feeding is usually done through a tube into the stomach or small intestine until the breathing tube is removed. Due to the

abnormal position of the stomach and esophagus, feeding difficulty is very common and can delay discharge.

What are the long term complications my baby may face?

Your baby may have long-term complications of CDH. These include:

- breathing problems
- recurrent lung infections
- gastrointestinal problems
- difficulty growing
- developmental problems

Local Resources:

For Further Information or Support:

Cherubs—Congenital Diaphragmatic Hernia
Support Group for parents and families of children with diaphragmatic hernia

<http://www.cherubs-cdh.org>

CHERUBS

Post Office Box 150

Creedmoor, NC 27522

919.693.8158 (telephone)

707.924.1114 (fax)

877.403.1944 (toll free) (U.S. only and for families who cannot afford long-distance calls)