CHOC Children's.

Diaphragmatic Hernia

A diaphragmatic hernia is a birth defect, which means it occurs before birth as a fetus is forming in the mother's uterus. The diaphragm has an abnormal opening, and with this type of birth defect, some of the organs that are normally found in the abdomen move up into the chest cavity through this opening.

There are two types of diaphragmatic hernia:

- Bochdalek hernia. A Bochdalek hernia usually involves an opening on the left side of the diaphragm. The stomach, liver, spleen and/or intestines usually move up into the chest cavity.
- Morgagni hernia. A Morgagni hernia involves an opening in the middle of the diaphragm close to the front of the chest.

What causes a diaphragmatic hernia?

As a fetus is growing in its mother's uterus before birth, different organ systems are developing and maturing. The diaphragm develops between the seventh and twelfth weeks of pregnancy. The esophagus (the tube that leads from the throat to the stomach), the stomach and the intestines are also developing at this time.

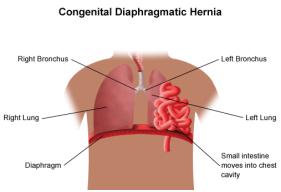
In a Bochdalek hernia, the diaphragm may not develop properly, or the intestine may become trapped in the chest cavity as the diaphragm is forming. In a Morgagni hernia, the tendon that should develop in the middle of the diaphragm does not develop properly. In both cases, normal development of the diaphragm and the digestive tract does not occur. Left-sided Bochdalek hernias make up about 80 to 90 percent of all cases. Morgagni hernias are much less common.

Why is a diaphragmatic hernia of concern?

The lungs are developing at the same time as the diaphragm and the digestive system. A diaphragmatic hernia allows abdominal organs to move into the chest cavity, instead of remaining in the abdomen as they are developing. With the heart, lungs and abdominal organs all taking up space in the chest cavity, the lungs do not have space to develop properly.

A diaphragmatic hernia is a life-threatening illness. When the lungs do not develop properly during pregnancy, it can be difficult for the baby to breathe after birth.

The intestines also may not develop properly, especially if they are not receiving enough blood supply while they are developing. A good blood supply is necessary for the intestines to develop correctly, and to be healthy and function properly.



What are the symptoms of a diaphragmatic hernia?

The symptoms of a Bochdalek diaphragmatic hernia are often observable soon after the baby is born. The following are the most common symptoms of a Bochdalek diaphragmatic hernia. However, each child may experience symptoms differently. Symptoms may include:

- Difficulty breathing
- Fast breathing
- Fast heart rate
- Cyanosis (blue color of the skin)
- Abnormal chest development, with one side being larger than the other
- Abdomen that appears caved in (concave)

A baby born with a Morgagni hernia may or may not show any symptoms.

How is a diaphragmatic hernia diagnosed?

Diaphragmatic hernia can often be detected on fetal ultrasound in the second and third trimesters of pregnancy. A fetal echocardiogram (ultrasound of the heart) may also be done to check for heart abnormalities before the baby is born.

If your baby has been diagnosed with a diaphragmatic hernia, we would be happy to schedule a consultation with your family and one of our top surgeons, as well as a CHOC neonatologist and your perinatologist, to prepare for the birth and subsequent care of your baby.

After birth, your baby's doctor will perform a physical examination. A chest X-ray is done to look at the abnormalities of the lungs, diaphragm and intestine. A blood test known as an arterial blood gas is often performed to evaluate the baby's breathing ability.

Other tests that may be performed include:

- Blood test for chromosomes (to determine if there is a genetic problem)
- Ultrasound of the heart (echocardiogram)

What is the treatment for a diaphragmatic hernia?

Treatment may include:

 Neonatal intensive care. A diaphragmatic hernia is a life-threatening illness and requires care in a neonatal intensive care unit (NICU). Babies with diaphragmatic hernias are often unable to breathe effectively on their own because their lungs are underdeveloped. Most babies will need to be placed on a breathing machine called a mechanical ventilator to help their breathing. CHOC Children's NICU is designated a Level 4 NICU—the highest level available because of the complex conditions we treat.

- ECMO (extracorporeal membrane oxygenation). Some infants may need to be placed on a temporary heart/lung bypass machine called ECMO if they have severe problems. ECMO does the job that the heart and lungs would be doing – putting oxygen in the bloodstream and pumping blood to the body. ECMO may be used temporarily while a baby's condition stabilizes and improves. CHOC is the only hospital in Orange County to offer ECMO.
- Surgery. When the baby's condition has improved, the diaphragmatic hernia will be repaired with an operation. The stomach, intestine and other abdominal organs are moved from the chest cavity back to the abdominal cavity. The hole in the diaphragm is repaired.

Many babies will need to remain in the NICU for a while after surgery. Although the abdominal organs are now in the right place, the lungs still remain underdeveloped. The baby will usually need to have breathing support for a period of time after the operation. Once the baby no longer needs help from a breathing machine (ventilator), he or she may still need oxygen and medications to help with breathing for weeks, months or years.

Can there be problems in the future?

Babies born with diaphragmatic hernias can have long-term problems and often need regular follow-up after going home from the hospital.

Many babies will have chronic lung disease and may require oxygen or medications to help their breathing for weeks, months or years.

Many babies will have gastroesophageal reflux. Acid and fluids from the stomach move up into the esophagus (the tube that leads from the throat to the stomach), and can cause heartburn, vomiting, feeding problems or lung problems. Gastroesophageal reflux can often be controlled with medications prescribed by your child's doctor.

Some babies will have difficulty growing. This is known as failure to thrive. The children with the most serious lung problems are most likely to have growing problems. Because of their illness, they often require more calories than a normal baby in order to grow and get healthier. Gastroesophageal reflux can also cause feeding problems, preventing a baby from eating enough to grow.

Some babies can have developmental problems. They may not roll over, sit, crawl, stand or walk at the same time healthy babies do. Physical therapy, speech therapy and occupational therapy are often helpful for these babies to gain muscle strength and coordination.

Some babies may have some degree of hearing loss. A hearing test should be performed prior to discharge from the hospital.

To schedule a consultation with a CHOC Children's pediatric surgeon, please call 714-364-4050.

Photo provided by Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities.