Congenital Diaphragmatic Hernia

Babies with congenital diaphragmatic hernia (CDH) are born with an opening in the diaphragm (a muscle that separates the lungs from the abdomen). The opening allows organs that are normally in the abdomen to enter the chest. This usually causes the lungs to develop abnormally: many babies with CDH develop breathing problems soon after birth. CDH can be a severe, life-threatening abnormality. Your baby will receive immediate evaluation and testing to find out how severe the problem is and to determine the best treatment.

What is congenital diaphragmatic hernia (CDH)?

Babies with CDH have an abnormal opening in the diaphragm. The diaphragm is a muscle, important for breathing, which separates the lungs from the abdomen. In CDH, some of the organs that should be in the abdomen go through ("herniate") the hole in the diaphragm, into the baby's chest. This can cause problems with your baby's lungs because they don't have enough room to develop normally. Other problems affecting the intestines and heart can also occur.

Some babies with CDH have a large hernia that causes serious breathing problems and other abnormalities. Others have a less severe hernia that causes fewer problems. Careful tests are needed to determine the severity and consequences of your baby's CDH. Special treatments may be needed to assist breathing for a period of time. After your baby's condition has stabilized, surgery can be done to repair the hole in the diaphragm.

What does it look like?

The medical problems of babies with CDH vary a lot, depending on the severity of the birth defect.

- Some babies with CDH develop severe breathing problems (respiratory distress) soon after birth. Less often, breathing problems develop some time after birth.
- If your infant's CDH is less severe, the symptoms may be different. Relatively mild breathing problems may occur. Vomiting or other digestive problems may result if the intestines become obstructed (blocked) in their abnormal position in the chest. In this condition, the intestines are not normally attached to the abdomen and can become twisted and obstructed (malrotation).
- Sometimes CDH is recognized before birth on a routine ultrasound scan.
- Some babies with CDH have other birth defects as well.

What causes CDH?

- CDH occurs when your baby's diaphragm develops with an abnormal opening. The opening allows some of the organs that should be in your child's abdomen to move into the chest. It is unknown what causes your child's diaphragm to develop in this way.
- Part or all of the stomach, intestine, spleen, and other organs can be found in the chest. This doesn't allow enough room for the lungs to develop normally. One lung, usually the left, is smaller than the other. The other lung may be squeezed to the side. Breathing problems occur because of the abnormally developed lungs and diaphragm.
- The lung abnormality leads to a problem called pulmonary hypertension—high blood pressure in the vessels carrying blood from the heart to the lungs. This can lead to further heart and lung problems.

What are some possible complications of CDH?

CDH is a serious birth defect. Modern treatments have greatly improved the survival rate. However, even with treatment, some infants with CDH die. The risk of death and severe complications is highest for infants with more severe CDH.

Infants who survive CDH may have medical problems related to poor lung development. Neurologic problems can also occur. The risk of these problems is higher in babies who need lifesaving extracorporeal membrane oxygenation (ECMO) treatment. Growth and nutrition problems are common as well.

Some infants with CDH need several operations or have to remain in the hospital for a long period of time for treatment and surgery.

What increases your child's risk of CDH?

- Congenital diaphragmatic hernia is a relatively common birth defect, affecting about 1 in 5000 infants. Many infants with CDH are stillborn (die in the womb).
- If anyone in your family had this birth defect, your child may be at higher risk. Otherwise, there are no known risk factors for CDH.

Can CDH be prevented?

There is no known way to prevent this birth defect.

How is CDH treated?

Infants with CDH need immediate evaluation and treatment. X-rays and other tests are performed to confirm that CDH is present and to determine how severe the abnormality is.

Treatment usually consists of two steps. Initial treatment focuses on *breathing support* to ensure that your baby is getting enough oxygen. Once his or her condition has stabilized, *surgery* is planned to move the organs into their proper position and to repair the hole in your baby's diaphragm.

- *Breathing support.* Treatments to help your baby breathe are the first priority. This often has to be done on an emergency basis in CDH babies who develop respiratory distress. Babies with CDH are treated in the intensive care unit (ICU), where they can receive constant treatment and round-the-clock monitoring.
  - Mechanical ventilation is often needed. Your baby will be connected to a machine called a ventilator to help the lungs perform the work of breathing.
  - In other infants, mechanical ventilation cannot provide enough oxygen. These infants receive a different kind of support called extracorporeal membrane oxygenation (ECMO). ECMO is a machine that takes over the functions of the lungs. It can keep your baby alive while his or her medical condition stabilizes. Your baby may need to stay on ECMO for a long time—2 weeks or even longer.
  - During either type of breathing support, your baby will receive constant medical monitoring and treatment. The goal is to allow time for your child to grow stronger before final repair of CDH is performed.
- *Surgery.* Babies with CDH eventually require surgery. More than one operation may be needed. The goals of surgery are:
  - To move the abnormally positioned organs back down to the abdomen.
  - To close the hole in your baby's diaphragm.
  - Surgery may be done as soon as 2 or 3 days after birth if your baby responds well to mechanical ventilation. If your baby requires ECMO, a delay of several weeks is more likely.

Every baby with CDH is different. Many factors affect your infant's medical condition, treatment, and chances of a good outcome.

When should I call your office?

Call our office if you have any questions about this birth defect or about your infant's treatment.

Please type your custom instructions and/or office contact information here.