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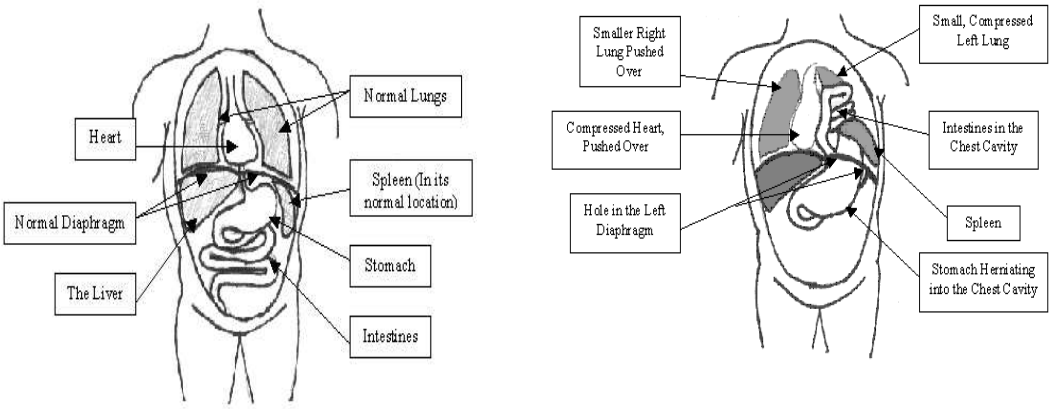
CONGENITAL DIAPHRAGMATIC HERNIA

What is Congenital Diaphragmatic Hernia?

The diaphragm is a thin dome-shaped muscle which separates the chest cavity (lungs and heart) from the abdominal cavity (intestines, stomach, liver, etc.). The diaphragm normally forms between the 7th and 10th week of pregnancy.
In babies with a Congenital Diaphragmatic Hernia (CDH), the diaphragm does not form completely. There is a hole in the diaphragm that allows part of the organs that are normally found in the abdomen to move up into the chest cavity.

This hole is found on the left side of the body about 85% of the time. It may also occur on the right side or in the middle.
A CDH can often be identified during a prenatal ultrasound, but some cases of CDH are not found until later in pregnancy or after birth.
Normal Anatomy Diaphragmatic Hernia



Normal Anatomy Diaphragmatic Hernia

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What are the causes of Congenital Diaphragmatic Hernia?

CDH occurs in about 1 in every 3000 births. Doctors do not yet fully understand what causes CDH. It appears to be a random event that can happen to anyone.

What does this mean?

A CDH allows parts of the intestines, stomach and/or liver to move into the chest cavity. These organs then put pressure on the lungs and heart. The lungs may not have space to develop properly. This is called pulmonary hypoplasia. The degree of pulmonary hypoplasia is usually one of the most important issues that

determine if the baby will be able to survive after birth. Babies that have a large hole in the diaphragm or have CDH diagnosed in early pregnancy can suffer from severe hypoplasia. These babies are less likely to survive after birth because they are unable to breathe.

The abdominal organs may also be affected. The intestines may not receive enough blood supply while they are developing to be healthy and able to function properly after birth. Other organs that can be affected are the kidneys, heart and the stomach.

So far, fetal surgery (fixing the CDH hernia while the baby is still in the mother's uterus) has not been proven to improve the health of the baby.
If other ultrasound or genetic problems are discovered there is a greater chance the baby will be affected mentally or physically. There is also a greater chance the baby will not survive. It is not possible to give you more information regarding your baby's health without further testing.

What other tests should we consider?

Approximately 40-50% of these babies will have another health problem, such as a defect of the brain or spinal cord such as spina bifida, a heart defect, a kidney problem, cleft lip/palate or a chromosome abnormality. There are also many other conditions that can be associated with CDH. It is important to check the baby for these other problems because it will help us tell you what it means for the development or survival of your baby.

Other tests may include a detailed ultrasound, a fetal heart ultrasound (echocardiogram), amniocentesis, and MRI. The detailed ultrasound is used to assess the baby's other organs. Magnetic resonance imaging (MRI) uses radiofrequency waves and a strong magnetic field rather than x-rays to provide detailed pictures of internal organs and tissues. There are no known harmful effects of MRI in pregnancy.

CDH is associated with chromosome abnormalities or problems such as Down Syndrome (trisomy 21). Some other genetic problems (trisomy 18) make it hard for babies to live longer than a few hours, days or months after birth and some genetic problems are so severe that babies are unable to survive after birth. You will be offered amniocentesis to look for genetic or chromosome problems. During an amniocentesis, a small amount of fluid is taken from the area around the baby and tested for these genetics problems. An amniocentesis may not find all genetic causes for CDH.

What will happen around the time of the baby's birth?

Even if the baby does not appear to have any other health problems, CDH is a life

threatening illness. Babies with CDH need to be born at a hospital with a neonatologist (specialist in newborn babies) and a pediatric surgeon (specialist in surgery of children). Babies with diaphragmatic hernia require care in a neonatal intensive care unit. They are often unable to breathe on their own and most need to be helped to breathe with a ventilator (breathing machine).

All babies born with CDH need surgery. It may take several days to weeks for the baby to be stable enough for surgery. At the time of surgery the abdominal organs will be moved from the chest cavity back in the abdominal cavity. The hole in the diaphragm will be fixed. After surgery, these babies need to remain in the NICU for a period of time.

Although the abdominal organs have been returned to the right place, the lungs still continue to be underdeveloped. The baby will need continued breathing support after surgery.

Babies with CDH can also have high blood pressure in the blood vessels going to the lungs. This is called pulmonary hypertension. This can cause the blood to back up and go around the lungs. Since less blood goes to the lungs it doesn't pick up as much oxygen and this can make the baby's oxygen levels very low. This is called shunting and can be hard to treat.

What does this mean for my baby's future?

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

Many babies will have long-term lung disease. They may need extra oxygen and medications to help them with their breathing.

Some babies have growing problems. They may need a feeding tube to give them the extra calories they need to grow and become healthier. Acid and fluids moving from the stomach into the esophagus can lead to heartburn, feeding problems, vomiting or lung problems.

Some babies may be delayed in development based on what is expected for a child of the same age i.e. they may not roll over, sit, crawl, walk, etc. at the same time as other children do. Delayed or damaged mental development has also been reported. They may need varied therapies to help them.

What do we do now?

You will meet with a doctor that specializes in high-risk obstetrics. The doctor will discuss with you in detail your options for further testing, discuss with you test results and provide you with treatment options. There is no right or wrong

answer on what to do next. Your doctor will provide you with all available information and assist you in making an informed decision. We will support you

in whatever decision you choose to make.

Where can I get more information?

Sick Kids Toronto
Respiratory Medicine: Congenital Diaphragmatic Hernia Clinic (CDH)
http://www.sickkids.ca/RespiratoryMedicine/What-we-do/Congential-diaphragmatic-hernia-clinic/index.html

The American Pediatric Surgical Association
Congenital Diaphragmatic Hernia
http://www.pediatricsurgerymd.org/AM/Template.cfm?Section=Home&TEMPLATE=/CM/ContentDisplay.cfm&CONTENTID=1359

The Children's Hospital of Philadelphia
Center for Fetal Diagnosis and Treatment: Congenital Diaphragmatic Hernia (CDH)
http://www.chop.edu/service/fetal-diagnosis-and-treatment/fetal-diagnoses/congenital-diaphragmatic-hernia-cdh.html