ROCKY MOUNTAIN PEDIATRIC SURGERY
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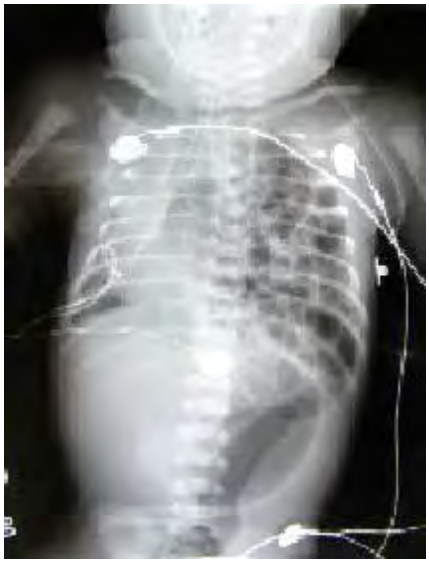
Congenital Diaphragmatic Hernia

What is a Congenital Diaphragmatic Hernia (CDH)?

One of the most challenging neonatal problems is congenital diaphragmatic hernia. A CDH occurs during the 10th week of development, when the diaphragm on one side, usually the left, fails to fully fuse closed. Abdominal contents then slip through the defect into the chest. Intestines, stomach, liver and spleen can slide up into the chest, compressing the heart and lungs. Although the presence of intestines in the chest sounds and appears dramatic, the mere presence of intestines in the chest is not in itself particularly dangerous. Instead, it is the abnormal pressure on the developing lungs and heart that cause problems when the baby is born. CDH is rare, occurring in about 1 in every 2000 to 1 in every 5000 live births. This relatively simple defect in the diaphragm disrupts cardiopulmonary (heart and lung) physiology, threatening the baby's life. Although medical therapy continues to improve, survival remains around 75%. No one knows exactly what causes CDH and mothers can do nothing to provoke or to prevent CDH.



This x-ray shows intestines (air inside them is black) in the chest and the heart is squeezed to the right side.

What Problems Can Arise from the Compression on the Developing Lungs?

- Decreased branching of airways resulting in fewer air sacs (alveoli) and therefore less surface area for gas exchange in the lung.
- A deficiency of surfactant, a substance that decreases the stiffness of the lungs
- Abnormally muscular and reactive blood vessels controlling flow of blood through the lungs, leading to high blood pressure in the lungs (pulmonary hypertension)

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These problems lead to an unfortunate downward spiral in that the small surface area of the lungs decreases oxygen content and increases carbon dioxide (CO_2) content in the blood. The low oxygen and high CO_2 cause abnormal blood vessels to squeeze, cutting down on blood flow to the lungs and causing the right heart to fail or to "shunt" blood around the lungs. The decreased blood flow to the lungs drops oxygen and raises carbon dioxide further. This cycle continues to repeat and if the cycle is not interrupted, the baby will die.

Not every baby with CDH falls into this cycle. Some babies are hardly affected, while others die before birth. But most require advanced medical care including careful mechanical ventilation, special inhaled and injectable drugs, sedation, and sometimes even a form of heart-lung machine called ECMO.

What Other Issues can arise from CDH?

- Defects in the heart
- Problems with the kidneys
- Side effects from aggressive medical treatment including lung damage or bleeding into the brain.

What is the Treatment for CDH?

- **Babies are exposed to less danger if they are born in a hospital with advanced high risk perinatology, neonatology, pediatric surgery and ECMO resources**. If the baby is born at a hospital without these services, the baby will have to endure a dangerous transport to a hospital that is equipped to handle such problems.
- **At birth, babies are placed on mechanical ventilation immediately**. While some babies will not need this, most do, and many can avoid the vicious cycle described above if high oxygen levels are maintained. Most babies have a "honeymoon period" of a few hours (6 – 24 hours) of relative stability and relatively low support. Then for unclear reasons, the baby suddenly deteriorates. A skillful intensivist can recognize this deterioration and take action to minimize the impact of these changes.
- **The main principle of care in these first days is to be patient:** It is known that the two main physiologic problems of low surface area and overly reactive blood vessels will improve if the baby can be supported through this period. The lungs will grow, adding air sacs and surface area, and the blood vessels will dilate and become less reactive. Whether these improvements will be enough to allow the baby to survive cannot be predicted. Nevertheless, these physiologic changes have important consequences for surgery.
- **In the past, it was believed that mechanical compression in the chest caused the abnormal physiology seen in these babies**. Emergency surgery to correct the defect and decompress the chest was thought to be advantageous to babies with CDH. It was later demonstrated that this reasoning was based on a misunderstanding of the physiological problem, and immediate emergency surgery actually decreased survival of CDH. For this reason, surgery is not performed until the baby has passed through the critical period and this time period is variable.
- **Traditionally, the diaphragm is repaired through an incision below the ribs on the side of the defect**. Often, there is enough diaphragm to repair the hole, but sometimes, a patch must be used to bridge the gap.
- **A minimally invasive surgical technique to repair CDH, usually through the chest (thoracoscopy) but occasionally through the abdomen (laparoscopy) can be performed in very stable babies**. Instead of a large incision, the repair is completed with 3 or 4 small (3 - 5mm) incisions, under vision by a fiberoptic camera. Not every baby is a candidate for a Minimally invasive repair. Your surgeon may discuss this option with you when the time comes.
- **Some groups have begun to experiment with fetal surgery techniques**. Although these techniques have produced predictable growth of the lungs, they have not resulted in improved cardiopulmonary physiology or survival. Furthermore, the method is risky to the baby and the mother, and remains experimental.

What is the Expected Outcome for my child?

Despite advances in medical and surgical technique, overall survival remains around 75%. It is helpful to think of three groups of babies with CDH: those who have little dysfunctional physiology; those with such severe physiological disruption that no intervention will allow survival; and, the largest group, those who need support and can be "recruited" to survive. Since the overall survival has changed little, we can surmise that

the group that will die despite any treatment has not changed in size. Instead, those recruitable babies tend to have fewer problems and long-term disability. Still, despite advances, some babies will have long term problems like reactive airway disease, right heart dysfunction or neurologic issues.

The severity of a baby's problems is difficult to predict before the baby is born. Some ultrasound measurements may suggest which babies will do poorly, but no measures reliably predict how sick a baby will be or the "probability of survival". While most babies survive, others do not, and some survivors have chronic diseases. Regardless of any "odds", your doctors and nurses know how to give your baby every advantage to gain the best possible outcome for your child.

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