

Patient Care > Congenital Diaphragmatic Hernia



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

One of the most challenging neonatal problems is congenital diaphragmatic hernia. Although rare (1 in 2000 to 1 in 5000 live births), the relatively simple defect disrupts cardiopulmonary physiology in peculiar ways, threatening the baby's life. Although medical therapy continues to improve, these improvements have been slow to improve outcomes for CDH; survival appears to remain around 75%.

Some mothers worry that they did something to cause or did not do something to prevent CDH. But no one knows exactly what causes CDH, and mothers can do nothing to provoke or to prevent CDH.

Anatomy and Physiology

CDH probably occurs during the 10th week of development, when the diaphragm on one side (usually the left) fails to fully fuse. 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.

Abnormal compression (and a possible genetic problem) on the developing lungs produces two distinct physiologic problems:

decreased branching of airways resulting in fewer air sacs (alveoli) and therefore less surface area for gas exchange in the lung (in fact, there is another problem, a deficiency of surfactant, a substance that decreases the stiffness of the lungs; but this only contributes to an inability to load oxygen and unload carbon dioxide).

abnormal sensitivity and strength of the blood vessels controlling flow of blood through the lungs, leading to pulmonary hypertension.

These two problems lead to an unfortunate downward spiral: small surface area decreases oxygen content and increases carbon dioxide content in the blood; low oxygen and high CO₂ cause abnormal blood vessels to squeeze, cutting down flow to the lungs and causing the right heart to fail or to “shunt” blood around the lungs; decreased flow to the lungs drops oxygen and raises carbon dioxide further; and the cycle continues. 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 even a form of heart-lung machine called ECMO (see link).

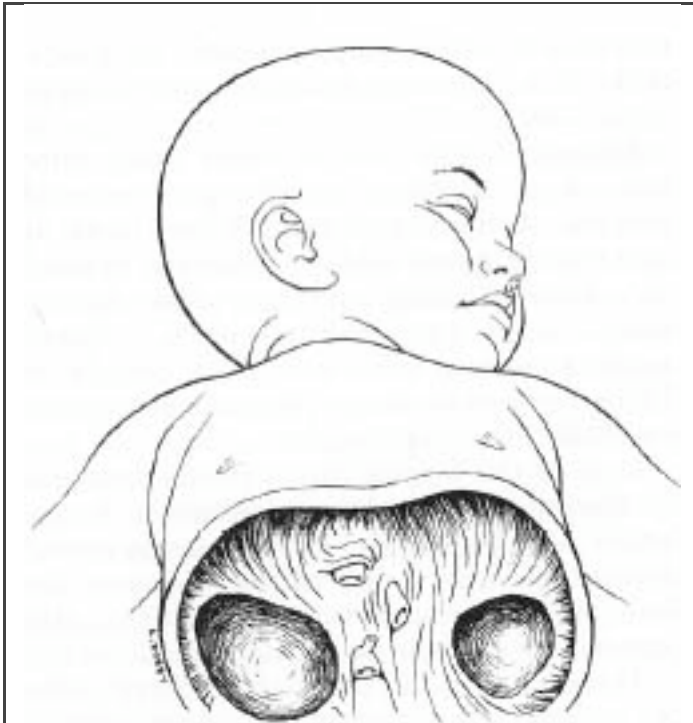
A few other problems may accompany CDH. The most common problems include defects in the heart, or problems with the kidneys. In addition, the baby may suffer side effects from aggressive medical treatment including lung damage or bleeding into the brain.

Treatment

Babies are exposed to less danger if they are born in the same hospital with advanced high risk perinatolgy, neonatology, pediatric surgery and ECMO resources. If the baby is born at a hospital without these, he will have to endure a dangerous transport to a hospital that is equipped this way.

At birth, babies are invariably 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) of relative stability and relatively low support. Then for unclear reasons, the baby will suddenly deteriorate. 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 gain time: It is known that the two main physiologic problems — low surface area and tight blood vessels — will partly correct 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.



Defects in the diaphragm may be on the right, left, or rarely, both sides.

Earlier, it was believed that mechanical compression in the chest caused the abnormal physiology seen in these babies. It followed that emergency surgery to correct the defect and decompress the chest should give the advantage to babies with CDH. Later it was demonstrated that this reasoning was based on a misunderstanding of the physiological problem, and emergency surgery actually decreased survival of CDH. For this reason, surgery is not performed until the baby has passed through the critical period.

Traditionally, the diaphragm is repaired through a large incision below the ribs on the side of the defect. Usually, there is enough diaphragm to repair directly, but occasionally, a patch must be used to bridge the gap. At RMPS, we use a collagen-based bio-membrane that allows the baby's native tissue to "grow in", decreasing the problems of foreign objects in the body and strengthening the repair.

At RMPS, we have pioneered a minimally invasive surgical technique to repair CDH. Instead of a large incision, the repair is completed with 4 or 5 small (3 - 5mm) incisions, under vision by a fiber optic digital camera. The approach is either through the abdomen or the chest. Not every baby is a candidate for a MIS repair. Your surgeon will discuss this option with you when the time comes. Still, our results with this technique have been good: the repair is mechanically the same as with the traditional technique, and some problems (like abdominal wall weakness, scarring and pain) are eliminated.

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.

Outcome

Despite advance 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, some babies will have long term problems like reactive airway disease and right heart dysfunction.

As mentioned, the severity of the baby's problems is difficult to predict before the baby is born. Some measurements can show which babies will do badly, 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.

Disclaimer: Your child's condition is unique. The information contained on this web site is not intended to substitute for advice from a doctor or nurse. If you are unsure about any aspect of your patient's care, please contact us at 303-839-6001, or talk to your pediatrician.