



Congenital Diaphragmatic Hernia

Background

Approximately one in 2,200 to 3,000 live births is affected by a congenital diaphragmatic hernia (CDH). This congenital defect allows abdominal organs to enter the thoracic cavity through a defect in the diaphragm during gestational development. CDH is a life-threatening condition, and prompt identification and management is needed in the delivery room because of the physical compression of the lung tissue on the affected side and the displacement of the heart and opposite lung within the thoracic cavity.

Definition

Approximately 85% of CDH defects occur in the left diaphragm, with defects occurring on the right or bilaterally much less frequently (Bradshaw, 2015). Of the left-sided defects, the majority occur in the posterior area of the diaphragm (Bochdalek hernia). Male newborns are more commonly affected. This defect carries a 2% recurrence risk in subsequent pregnancies (Carley, 2007). The severity of the defect is related to the timing and degree of herniation of abdominal contents during fetal development.

Some defects are very small and can be difficult to identify. Others are large, even progressing to complete absence of the diaphragm. Larger defects that herniate into the thoracic cavity have a higher incidence of bilateral pulmonary hypoplasia, persistent pulmonary hypertension, and intrapulmonary shunts. The abdominal contents that have herniated into the thoracic cavity compress the lung tissue, limiting the lung's growth and ability to oxygenate and ventilate. This pulmonary compression causes marked diminution of bronchial branching, limited multiplication of alveoli, and the persistence of muscular hypertrophy in the pulmonary arterioles. These abnormalities occur on the affected side, but they may affect the "nondefect" side as well, usually to a lesser degree. The exact etiology for CDH is unknown, and it may present as an isolated defect or as part of a syndrome.

Associated Conditions

Associated anomalies occur in more than 40% of infants with a CDH. The most common anomalies involve the central nervous, cardiovascular, musculoskeletal, gastrointestinal, and genitourinary systems. CDH also has been noted to be a component of chromosomal abnormalities such as trisomies 13, 18, and 21.

Diagnosis

CDH commonly is diagnosed on prenatal ultrasound. A common maternal association is polyhydramnios. The hallmark finding on prenatal ultrasound is a fluid-filled stomach behind the left atrium. However, CDH is not always diagnosed prenatally due to the size of the defect and the positioning of the fetus during the ultrasound evaluation.

Clinical Presentation

These infants often will present with significant respiratory distress and cyanosis in the delivery room or within the first few minutes of life. The amount of work it takes for them to breathe typically gets worse over time. If the infant received bag and mask ventilation, their condition will significantly worsen because the ventilation inflates the bowel along with the lungs. As the bowel becomes distended from the ventilation, it further compresses the lung tissue. Other signs include a scaphoid abdomen, barrel chest, heart sounds shifted from the normal point of maximal impulse location, decreased breath sounds, signs of decreased cardiac output such as poor perfusion, bowel sounds auscultated in the chest, and decreased oxygen saturations. The hypoxemia noted in these infants often is related to right to left shunting through their ductus arteriosus, foramen ovale, and intrapulmonary shunts.

Laboratory Findings

It is common to obtain an arterial blood gas on a patient with this degree of respiratory distress and cyanosis. The blood gas will often reveal hypoxemia, hypercapnia, and a metabolic and/or respiratory acidosis.



On chest X ray, the gastric bubble may be displaced. This is not always the case, depending on whether the stomach has migrated to above the diaphragm. Typically, the bowel gas pattern is noted in the thorax. This bowel gas may not be seen immediately after birth.

Delivery Room Management

Ideally, if the baby's defect is identified prior to birth, the mother will deliver in a center capable of providing inhaled nitric oxide, extra corporeal membrane oxygenation (ECMO), pediatric surgery, and genetic evaluation to the infant. Late preterm and term infants who present with significant respiratory distress and cyanosis in the delivery room should be evaluated for a CDH. These infants do not respond well to bag and mask ventilation for reasons addressed above.

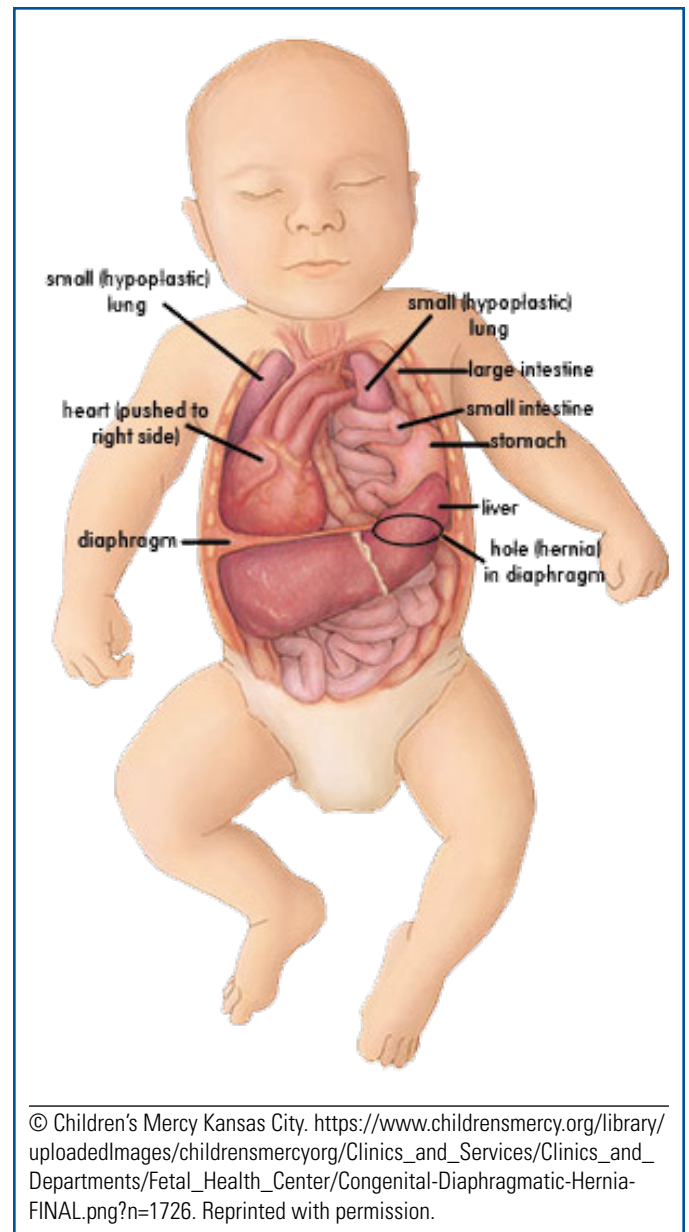
For infants with a known CDH, immediate endotracheal intubation and ventilation is done to minimize the overdistention of the stomach and intestines. This overdistention could lead to further compression of the lung tissue. To further improve the ability to ventilate and oxygenate this newborn, a Replogle or orogastric (OG) tube should be inserted. Another priority in the immediate care of this newborn is to minimize pulmonary hypertension (see *Persistent Pulmonary Hypertension in the Neonate* in Diagnoses) by evaluating and treating systemic hypotension, respiratory acidosis, and metabolic acidosis.

Preoperative Care

CDH is no longer considered a surgical emergency. Stabilization of this newborn is very important. This extra time also allows for thorough evaluation of any associated conditions that may preclude ECMO or surgery.

From a respiratory standpoint, the baby will be intubated and receive mechanical ventilation. Avoid bag or mask ventilation whenever possible. It also is important to evaluate for any signs of a pneumothorax. The pulmonary hypoplasia makes the newborn's lungs less compliant and more at risk for a pneumothorax. For these reasons, many babies with CDH will be managed with high-frequency ventilation. Other important aspects of care

include maintaining adequate perfusion, identifying and correcting any acid base imbalances, maintaining adequate oxygenation, and administering surfactant. If mechanical ventilation is not effective, the baby may require escalation of care, including inhaled nitric oxide or ECMO. If not already done, a Replogle or OG tube should be inserted to decompress the bowel. Central vascular access (umbilical lines or peripherally inserted central catheter and peripheral intravenous [IV] lines) will be needed for IV fluid and medication administration as well as arterial blood-pressure monitoring and frequent blood draws.





Surgical Care

At this time, in utero repair of the CDH does not seem to improve overall survival when compared with the standard postnatal repair (Gomella, 2013). Usually, a primary closure is possible. Synthetic patches may be used in the repair of large defects or when the diaphragm is absent.

Postoperative Care

Important aspects of postoperative care for a baby with CDH include pain management, maintaining adequate ventilation and oxygenation, continued gastric decompression, fluid and electrolyte management, and continued monitoring. The baby may be restricted to nothing by mouth for several days, so parenteral nutrition will be important for this baby. If right-to-left shunting is a problem, then inotropes (dopamine or dobutamine) may be used to decrease this shunting. Chest tubes (see *Chest Tubes* in Procedures) always should be placed to gravity or water seal; suction is avoided to prevent any acute mediastinal shifts.

Long-Term Follow-Up and Prognosis

Survival rates vary depending on the size of the defect, the amount of herniation (bowel in the chest), lung development, the presence of pulmonary hypertension, and any other associated conditions. As with other

congenital defects, early intervention has been shown to help improve outcomes. The mortality for newborns with CDH is approximately 50%. Follow-up evaluation is an essential component of the care of babies with CDH. It has been noted that some survivors have abnormal tone and delayed neurocognitive and language skills.

References

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Congenital Diaphragmatic Hernia: Information for Parents

Around one in 2,200 to 3,000 newborns is born with a congenital diaphragmatic hernia (CDH). The diaphragm is the muscle that separates the chest cavity from the abdominal cavity. It is a very important part of breathing normally. This defect allows the stomach organs to enter the space in the chest where the baby's heart and lungs are. When this happens while the baby is still developing, it can be a life-threatening condition. It is important that the medical team identify this condition as soon as possible.

Most CDHs occur on the baby's left side; male babies tend to have them more often. Some CDHs are very small and can be difficult to find. Others are large, sometimes with a complete absence of the diaphragm. Because the stomach contents are inside the area where the heart and lungs are, the normal growth of the heart and lungs may be affected.

Many babies with CDH have other problems. The most common problems involve the brain or spinal cord, the heart, the bones or muscles, the intestines, or the kidneys and bladder. Babies with CDH also may have issues with their chromosomes.

Although CDH is commonly diagnosed during a prenatal ultrasound, it is not always found before delivery. Ideally, if your baby's CDH is known before birth, you should deliver in a hospital with a level III or IV neonatal intensive care unit so that your baby will receive the care he or she needs right away. If not diagnosed before delivery, medium to large CDH defects will be diagnosed after birth. If this happens, your baby may need to be moved to another hospital. Often these babies will look blue in the delivery room, or within a few minutes after

birth, and will have a hard time breathing. You may notice that their chests are big and their stomachs are sunken in. There may be signs that their hearts are having trouble pumping blood to the body (weak pulses and pale skin). These babies usually require a breathing tube to be placed in their windpipes to make breathing easier.

A soft plastic tube will be placed in your baby's mouth to remove air from his or her stomach. This will help get more oxygen to your baby. He or she will not be fed for several days. An intravenous (IV) catheter will need to be placed to provide fluids, nutrition, and medications.

When your baby is stable, he or she will go to surgery to correct the CDH. Depending on the size, your baby may require heart-lung bypass before or after surgery. The heart-lung bypass machine helps do the job of the lungs and heart to allow them to heal.

Most times, the CDH can be repaired with one surgery. After surgery, the baby's care will include receiving baby pain medicine and nutrition and medication through an IV. Your baby may have a chest tube (see *Chest Tubes in Diagnoses*). This is a soft plastic tube that removes the air from the area outside of the baby's lungs. This will be taken out at the bedside a few days after surgery.

Survival rates vary depending on the size of the CDH and the complications both before and after surgery. Unfortunately, about 50% of babies do not survive. Follow-up appointments are very important in the care of babies with CDH. Some babies who survive can have abnormal muscle tone and delayed neurocognitive and language skills.