Congenital Diaphragmatic Hernia and Diaphragmatic Eventration

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KEYWORDS

• Congenital diaphragmatic hernia • CDH • Morgagni hernia • Bochdalek hernia

KEY POINTS

- Congenital diaphragmatic hernia (CDH) occurs in approximately 1 in 2500 to 5000 children.
- The lack of uniform early diagnosis and referral has led to discrepancies in reporting incidence and survival, and resulted in the "hidden mortality."
- Two main types of CDH occur: Morgagni (anteromedial) and Bochdalek (posterolateral).
- Bochdalek hernias occur at a much greater frequency, and are typically associated with physiologic derangements in the newborn.

INTRODUCTION: NATURE OF THE PROBLEM

Congenital diaphragmatic hernia (CDH) occurs in approximately 1 in 2500 to 5000 children. The lack of uniform early diagnosis and referral has led to discrepancies in reporting incidence and survival, and resulted in "hidden mortality," a term coined by Dr Michael Harrison in 1978. Two main types of CDH occur: Morgagni (anteromedial) and Bochdalek (posterolateral). Because Bochdalek hernias occur with much higher frequency, and are typically associated with physiologic derangements in the newborn, they are the focus of this article. Repair of the hernia can take many forms, with an open or minimally invasive technique, with or without a patch, and approached from the abdomen or thorax. The attendant pulmonary hypertension and hypoplasia experienced by patients with CDH are the main determinants of survival, and therefore the focus of perinatal management.

INDICATIONS AND CONTRAINDICATIONS

Congenital anomalies of the diaphragm span the range of disease severity, from an asymptomatic mild eventration resulting from shoulder dystocia during childbirth to

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complete absence of the diaphragm in the setting of a severe Bochdalek hernia. The indication for surgery in diaphragmatic hernia patients is straightforward—the presence of a hernia necessitates repair almost universally. The exception to this is an infant who is so profoundly unstable with no chance of recovery or adaptation of the pulmonary pressures.

SURGICAL TECHNIQUE AND PROCEDURE

Preoperative Planning

Management of the patient with CDH before surgery can take several different forms in terms of repair timing, but all hold several basic tenets in place: permissive hypercapnia (target range of $Paco_2$, 45–60 mm Hg), maintenance of preductal oxygen saturation (goal of 85%–95%), minimize volutrauma and barotrauma with conventional ventilator strategies aimed at keeping a positive inspiratory pressure of less than 25 cm H_2O with a positive end-expiratory pressure of between 2 and 5 cm H_2O. If this cannot be accomplished, conversion to high-frequency oscillatory ventilation is used, optimizing alveolar recruitment with a mean arterial pressure of 13 to 17.[1–3] The addition of inhaled nitric oxide can further decompress the pulmonary vascular bed. Pressors may be necessary, aimed at driving up inotropy. If these goals cannot be accomplished using maximal medical therapy, extracorporeal membrane oxygenation (ECMO) is required to deliver oxygen and remove CO_2. The typical indication for ECMO is a sustained oxygen index of greater than 35 to 40. At our institution, we typically await resolution of increased pulmonary artery pressures before proceeding with repair. These signs include decreased pulmonary artery pressures (using preductal arterial oxygen saturation as a surrogate marker, indicating a low degree of right-to-left shunting), along with decreased pressor requirement, and echocardiography showing decreased signs of right ventricular strain.[4] Use of ECMO beyond 4 weeks duration has been disputed.[5] At other institutions, the CDH is repaired early while still on ECMO.[6] Blood products are prepared in advance for the almost certain need for perioperative transfusion.

Preparation and Patient Positioning

The infant is laid across the operating table at the end of the bed such that the face is directed toward the anesthesiologist. The patient is placed in the lateral decubitus position with the affected side up, and the body prepped from the shoulder to the pelvis, including the abdomen. The head is placed at a slightly obtuse angle leaning toward the unaffected side, beyond midline, so as to minimize instrument strike while working thoracoscopically. An axillary roll is placed to protect the brachial plexus (depending on patient size, this may be a gel roll or soft gauze), and the patient is secured to the bed. An additional roll is placed to support the back. The surgeon and assistant stand at the head of the bed and a screen is positioned near the patient's feet. In the event that conversion to an open procedure is necessary, the tape securing the baby is split by a nurse, maintaining sterility, and the roll removed from behind the back.

Surgical Approach

These authors prefer a thoracoscopic approach for both CDH and eventration, regardless of the side. This allows for a better suturing angle and facilitates reduction of the viscera in a CDH.[7] We use the thoracoscopic approach for any infant that is stable enough to be transported to the operating room. If the infant is requiring high ventilator support or is on ECMO, and hence too unstable to be transported, we perform an open repair in the neonatal intensive care unit.