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Congenital Diaphragmatic Hernia: Management & Outcomes

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Keywords

• Congenital diaphragmatic hernia • CDH outcomes
• CDH pulmonary hypertension • CDH ventilation • CDH ECMO

Key points

- The successful management of congenital diaphragmatic hernia is truly a multidisciplinary team composed of expert neonatologists and pediatric surgeons.
- Pulmonary hypertension and lung hypoplasia are primary drivers of neonatal CDH morbidity and mortality. Initial treatment in the postnatal period is aimed at minimizing lung trauma and decreasing pulmonary hypertension crises.
- Surgical repair of congenital diaphragmatic hernia is not urgent in most circumstances and can be delayed until the pulmonary status of the patient has stabilized.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a congenital defect of the diaphragm in about 1:3000 live births that leads to a lack of separation of the abdominal and thoracic cavities during critical stages of development. The movement of abdominal contents in the thoracic space during this essential time can cause pulmonary hypoplasia from the mass effect of those abdominal contents. Not only is the lung small but it also lacks a normal bronchial and vascular branching pattern. This abnormal development leads to increased pulmonary vascular resistance and resultant significant pulmonary hypertension. It is this abnormal lung development and resultant pulmonary hypertension that is the major source of morbidity and mortality in infants with CDH [1,2].

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Successful management of the postnatal pulmonary hypertension is the key factor in survival of these children. Given the small patient population size it is challenging to do comprehensive prospective randomized trials to develop clear evidenced-based guidelines for management, although there now is decades worth of experience in modern prenatal imaging, intensive care unit management, and surgical techniques to draw on in creating expert opinion–based guidelines [3,4].

There are two major types of CDH: Bochdalek and Morgagni. Morgagni hernias, which are often incidentally discovered in older children, are a defect in the central anterior portion of the diaphragm and rarely lead to the mass effect and lung maldevelopment. The term Bochdalek hernia refers to what is classically thought of as a CDH in infants, a defect in the posterior lateral portion of the diaphragm. It is Bochdalek hernias and their management and outcomes that we refer to in this article.

The advances in neonatal intensive care have improved the survival of patients with CDH but this has also left a population of patients with significant morbidities. In this review we also discuss the outcomes of CDH management.

PRENATAL DIAGNOSIS

One of the largest advances in the care of the CDH patients in the modern era comes from the advent of improved prenatal imaging and access to that imaging. More than two-thirds of CDHs patients in the United States are now diagnosed prenatally. The most common form of CDH, the left-sided defect, with abdominal contents herniating into the thorax is often easily detected by ultrasound. Many fetal centers proceed to fetal MRI and fetal echocardiography after the detection of a potential diaphragm defect on ultrasound. Sometimes the findings of CDH are subtle, such as abdominal positioning of mediastinal structures or pleural effusion [4,5].

Adequate counseling of parents requires the ability to give them some predictive information based on the prenatal assessment. Many different predictors have been suggested. Herniation of the liver into the left chest is known to be poor prognostic factor but is challenging to qualify. If there is liver herniation into the chest this often suggests a large structural defect in the diaphragm thus also suggesting more impact on the thoracic organs and worse pulmonary hypoplasia and pulmonary hypertension. Hedrick and colleagues [6] demonstrated that liver herniation is highly predictive of the need for extracorporeal membrane oxygenation (ECMO) and also predictive of increased mortality (65% with herniation, 7% without herniation).

Observed-to-expected lung-head ratio (O/E LHR) has become one of the standard prenatal metrics to help predict morbidity and mortality. LHR is the ratio of the right lung area to the head circumference. Adding an O/E calculation helps correct for the rapid growth in lung development in third trimester. Combing O/E LHR and liver herniation helps estimate a survival percentage but ultrasound along with some newer MRI techniques has the