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MINI REVIEW

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Diaphragmatic hernia in children: A mini-review

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Abstract

The condition known as congenital diaphragmatic hernia (CDH) is an extremely uncommon birth defect that is characterized by incomplete closure of the diaphragm and herniation of fetal abdominal organs into the chest. This leads to pulmonary hypoplasia, postnatal pulmonary hypertension due to vascular remodeling, and cardiac dysfunction. The severity of cardiopulmonary pathophysiology is directly related to the high mortality and morbidity rates that are associated with CDH. Even though the etiology is still a mystery, it is believed that approximately one-third of cases of CDH have a polygenic origin. Antenatal ultrasonography, fetal magnetic resonance imaging (MRI), echocardiography, and other diagnostic procedures are typically used in conjunction with antenatal ultrasound to diagnose CDH. Prenatal management can include fetal endoscopic tracheal occlusion, which is a surgical intervention aimed at promoting lung growth in utero. This type of treatment is only available at specialized centers. In severe cases, postnatal management may involve extracorporeal life support in addition to the standard focus on cardiopulmonary stabilization. The rapidly shifting landscape of therapeutic options, which includes the management of pulmonary hypertension, various ventilation strategies, and surgical approaches, is one of the primary factors driving the ongoing development of clinical practice guidelines. Survivors frequently suffer from long-term morbidities that affect multiple body systems, such as pulmonary dysfunction, gastroesophageal reflux disease, musculoskeletal deformities, and impaired neurodevelopment. Research that is just getting started focuses on small RNA species as potential severity biomarkers and regenerative medicine approaches to improve the development of fetal lungs.

Introduction

Diaphragm discontinuity is caused by congenital diaphragmatic hernia (CDH), a developmental closure defect. Viscera from the abdomen can herniate into the chest as a result. A high rate of neonatal morbidity and mortality is caused by CDH, which affects one in every three thousand live births (1). Additionally, it has been linked to pulmonary hypertension and severe pulmonary hypoplasia. The defect is most frequently located on the posterolateral left side of the diaphragm (75–90% of cases), but it can also be right-sided (10–15%) or even bilateral (1–2% of cases) (2). According to some authors, male fetuses have a slightly higher incidence of CDH. The prevalence of CDH does not seem to be related to the age of the mother. In utero herniation of the viscera may cause frequent fatal complications, despite the fact that diaphragmatic herniation is surgically treatable. The proper patient selection and eligibility for the FETO (fetal endoscopic tracheal occlusion) surgery are crucial now that it is available. To minimize pulmonary hypoplasia and lower mortality, FETO is used. Like any surgical procedure, FETO has some limitations and poses a risk of unfavorable side effects (3).

Epidemiology

The prevalence of CDH is thought to be 2.3–2.4 per 10,000 live births in Europe and the US, and it has shown a slight but significant rise over time. With CDH, a high percentage of fetuses experience termination or stillbirth, which is frequently accompanied by other congenital abnormalities. The prevalence of CDH is likely underestimated overall because 25–35% of fetuses with prenatal CDH diagnoses end up having their pregnancy terminated, passing away in utero, or passing away soon after birth. As a result, many newborns with CDH that was diagnosed during pregnancy might never be identified or treated in a tertiary referral facility. These patients, who are thought to have the most severe CDH of any CDH infants, add to the "hidden mortality" of CDH (1,3).

Male infants are more frequently affected by CDH, and left-sided CDH accounts for the majority of cases (80%), followed by right-sided CDH (19%) and bilateral CDH (1%). Ninety percent of all CDH cases are found posterolaterally, or "Bochdalek," and the remaining cases are anteriorly, where they are known as "Morgagni" hernias and also include defects of the central septum transversum. A much worse prognosis is predicted by bilateral diaphragmatic hernias, which are more frequently linked to other congenital anomalies. Recent epidemiological research has not found any correlation between CDH and maternal age (4).

Prenatal diagnosis

Prenatal counseling, patient triage, and the identification of high-risk infants with CDH all benefit from accurate prenatal diagnosis and prognostication of disease severity. It's crucial to distinguish CDH from other intrathoracic anomalies, where normal anatomy is otherwise unaltered, in order to make an accurate diagnosis. These include mediastinal lesions such as enteric, neurenteric, or thymic cysts, as well as CPAMs, bronchogenic cysts, bronchial atresia, or bronchopulmonary sequestrations. The differential diagnoses must also take into account diaphragm eventration. Even though it can be difficult to tell eventration from CDH, it has a better outlook and a different management strategy. Even though eventrations are typically isolated lesions, they can become complicated by pericardial and/or pleural effusions. Between 50 and 70 percent of newborns with CDH are discovered during pregnancy, and over the past two decades, prenatal detection has become much more common. Between the 18th and 22nd weeks of pregnancy, ultrasound (US) screening exams are most frequently used to diagnose CDH (5).

Polyhydramnios, intrathoracic fluid-filled bowel loops, an echogenic chest mass, mediastinal shift, and/or an intrathoracic stomach are among the fetal US features (6,7). Left-sided CDHs are more frequently found before birth and include herniation of the stomach, intestines, and/or spleen as well as mediastinal/cardiac shift to the right. Although the liver can herniate, it can be more challenging to distinguish because of how often it echoes with the lung. The right lobe of the liver is herniated in right-sided CDH and the mediastinum shifts to the left side (6,8).

Clinical presentations

Typically, newborns with CDH exhibit respiratory distress. The clinical spectrum at birth includes a newborn with no symptoms, immediate, profound respiratory distress, respiratory acidosis, and hemodynamic instability, as well as an initial stable period with delayed respiratory distress. The initial symptoms of respiratory distress include tachypnea, retractions of the chest wall, grunting, cyanosis, and/or pallor. Infants often have a scaphoid abdomen and may show a slight increase in thoracic diameter upon physical examination. A physical finding associated with mediastinal shift is the frequent displacement of the point of maximal cardiac impulse. As breath sounds decrease bilaterally, bowel sounds may be audible in the thoracic cavity. Chest excursion could be lessened, which would indicate a lower tidal volume (9).

A chest radiograph that shows intestinal loops within the hemithorax, cephalad displacement of the stomach and orogastric tube and a mediastinal shift in the direction of the opposite hemithorax is typically used to confirm the diagnosis of CDH (Figure 1). In the beginning, there may be little to no gas in the abdominal cavity. CDH on the right side can be difficult to diagnose. Salient features, such as intestinal and gastric herniation, may not be obvious, and the right lobe of the liver that has herniated may be mistaken for an eventration or elevation of the right diaphragm (10). When lung compression features are the only radiographic sign, they can be mistaken for cystic teratomas, CPAMs, pulmonary sequestrations, bronchopulmonary cysts, neurogenic cysts, or bronchopulmonary cysts. Even though 20% of newborns with CDH may present outside the neonatal period, the majority of them will be identified within the first 24 hours of life. These patients have gastrointestinal pathology, feeding intolerance, chronic pulmonary infections, pleural effusions, pneumonia, and mild respiratory symptoms. Some children may present with intestinal obstruction or volvulus because CDH is invariably associated with abnormal intestinal rotation and fixation (11).

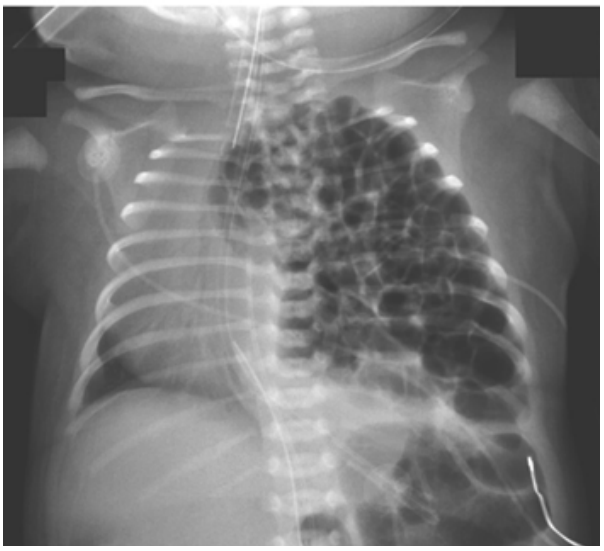

Figure 1: Chest radiograph

Operative repair

The surgical repair of the diaphragmatic defect in CDH can be done in one of two ways: open or minimally invasive. There are numerous subcategories that fall under these broad categories. In a recent analysis of CDHSG data, 84% of repairs were open, and of these, 95% were performed via laparotomy (96% of laparotomies are performed via a subcostal incision) (12).

Open approach

An abdominal or thoracic approach can be used to perform an open repair of CDH. The ability to mobilize the posterior rim of the diaphragm, easier management of intestinal rotational anomalies (if necessary), and avoidance of thoracotomy-associated musculoskeletal sequelae are all benefits of laparotomy (13). With a few centers choosing to use a midline laparotomy incision and the remaining centers performing thoracotomies, subcostal incisions account for the vast majority (>90%) of open neonatal repairs for CDH. With great care, the herniated contents should be removed from the hemithorax. It is necessary to remove a true hernia sac, which occurs less frequently than 20% of the time. Pulmonary sequestration should be looked for in the thoracic and abdominal cavities (14,15).

Even with this "gold standard" abdominal approach, open CDH repair's respiratory consequences and morbidity are still a concern. Despite the enlarged thoracic space created in the ipsilateral hemithorax, breathing compliance can be significantly decreased after open repair in addition to pulmonary hypoplasia and hypertension. When compliance falls by more than 50%, which can happen as a result of a tight abdominal wall closure, mortality increases significantly (16,17). Peak airway pressures should be carefully monitored as the abdominal fascia closes. This risk can be reduced by using a broad patch that bends into the hemithorax. The surgeon should think about a relaxed, temporary abdominal closure (fascial patch/prosthetic silo, vacuum-assisted closure, or skin-only closure), with planned delayed fascial closure, which is used in about 10% of cases, if respiratory compromise results. With right-sided CDH and infants on ECMO, this method is more frequently necessary. Delayed closure should be tried after the generalized edema has subsided or the intra-abdominal domain has expanded, especially in infants receiving ECMO (16).

Minimally invasive approach

Many surgeons have adopted minimally invasive surgical (MIS) approaches to CDH repair in an effort to prevent respiratory sequelae and other morbidity seen after open repair. This is due to the advancement of surgical techniques and the optimization of perioperative respiratory care. According to data from the CDHSG, laparoscopic and thoracoscopic techniques have been used in 16% of all CDH repairs in the most recent period. Primary repair and prosthetic patch closure have both been done using MIS techniques, which may have the benefits of less postoperative pain (4,7,8,17).

Conclusions

Congenital Diaphragmatic hernia is a complex abnormality. If the defect is severe and the prognostic factors are combined, the prognosis still remains very poor. Despite modern and intensive care, mortality and morbidity rates continue to be high. The most serious neonatal complications are pulmonary hypertension and lung hypoplasia. Different levels of bilateral pulmonary hypoplasia could account for the severity differences between neonates exhibiting respiratory distress and CDH.

Conflict of interest

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