ISSN 2598-3180 (Online)
JOURNAL OF MIDWIFERY
Research and Practice
Accredited by SINTA 4th

Article

Antenatal Ultrasound Should Be For All; Congenital Diaphragmatic Hernia - A Case Report

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SUBMISSION TRACK

Recieved: 14 October 2021
Final Revision: 26 Novemver 2021
Available Online: 27 Desember 2021

KEYWORDS

Hernia, Diaphragmatic, Prenatal Care, Newborn

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Abstract.

Congenital diaphragmatic hernia is an anatomical defect of the diaphragm, which allows protrusion of abdominal viscera into the chest, causing serious pulmonary and cardiac complications in the neonate. In this study we aimed to present a case of congenital diaphragmatic hernia. We investigated a 35-36 weeks of pregnancy, with a pregnancy carried out in a public hospital in Padang, West Sumatera. We suggest that if diagnosis occurs in the prenatal period, the prognosis of this disease improves.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) has an incidence of 1 per 2,000 to 3,500 deliveries. Left-sided CDH commonly contain stomach, bowel, and spleen and is associated with a generalized pulmonary hypoplasia.(1) The clinical presentation depends on the extent of pulmonary hypoplasia and may vary from cyanosis and respiratory distress immediately after birth to late presentation of respiratory distress in the first few days of life when the herniated bowel loop fills with air and compresses the ipsilateral lung parenchyma.(2) Typical signs include abnormally sited cardiac impulse, absent breath sounds, and scaphoid abdomen.

The diaphragm is a dome shaped musculotendinous partition that separates the thoracic cavity from peritoneal cavity(3). Its development begins at approximately 3 weeks of intrauterine life and completes by 9^{th} week. It is formed by fusion of four developmental compartments: the ventral or septum transversus, the dorsal esophageal mesentery, the pleuroperitoneal membrane and lateral aspects formed from muscular components of the body wall.

The most common type is the defect in closure of pleuroperitoneal membrane known as Bochdaleck hernia accounting for 90% of all CDH. It usually occurs as posterolateral defect affecting the left hemi thorax in most of the cases. When present in newborn it is frequently associated with significant respiratory insufficiency resulting in mortality depending upon varying degrees of pulmonary hypoplasia(4).

The diagnosis is commonly made on prenatal ultrasound and confirmed on postnatal chest radiography. Medical management consists of initial resuscitation with a view to avoid face mask ventilation, preoperative stabilization, and ventilatory care aiming in reducing the risk of volutrauma and persistent pulmonary hypertension of the newborn, while the surgical treatment consists of reduction of the herniated contents and closure of the diaphragmatic defect.

Congenital diaphragmatic hernias are often classified by their position. A Bochdalek hernia is a defect in the side or back of the diaphragm (Figure.1). Between 80 and 90 percent of congenital diaphragmatic hernias are of this type. A Morgnani hernia is a defect involving the front part of the diaphragm. This type of congenital diaphragmatic hernia, which accounts for approximately 2 percent of cases, is less likely to cause severe symptoms at birth. Other types of congenital diaphragmatic hernia, such as those affecting the central region of the diaphragm, or those in which the diaphragm muscle is absent with only a thin membrane in its place, are rare(7,8,9)

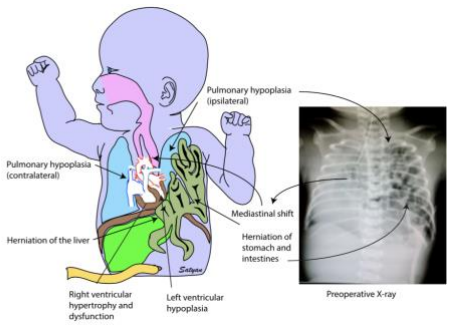

Figure.1

METHODS.

Case Report According to our case report, we believe that it is helpful in terms of providing neonatologists, surgeons and maternal-fetal medicine specialists with realistic prognostic information for counseling families. Nonetheless, several cautions are worth noting. Furthermore, it was recently established a multidisciplinary CDH clinic that was composed by specialists from a variety of areas, including pediatric general surgery, pulmonary, cardiology and nutrition [2]. This clinic allows us to make a more in-depth evaluation of the CDH and it certainly allows for better follow-up of children with this disease in a prospective mode.

RESULT.

We investigated a male patient, born at 35-36 weeks by C-Section on indication IUGR, weighing 1365 g, Body length 38 cm, with Apgar score 6/7 in the first and fifth minutes of life, which required positive pressure ventilation (PPV) at birth. His mother make complete prenatal care with OBGYN and have ultrasonographic examination of the current pregnancy demonstrated Intra uterin growth restrictin At 32 weeks of gestation. A repeat ultrasound scan at 35-36 weeks of gestation in adequaternary level fetal medicine unit showed Intra uterin growth restrictin with left sided congenital diaphragmatic hernia(CDH) (Figure.2)

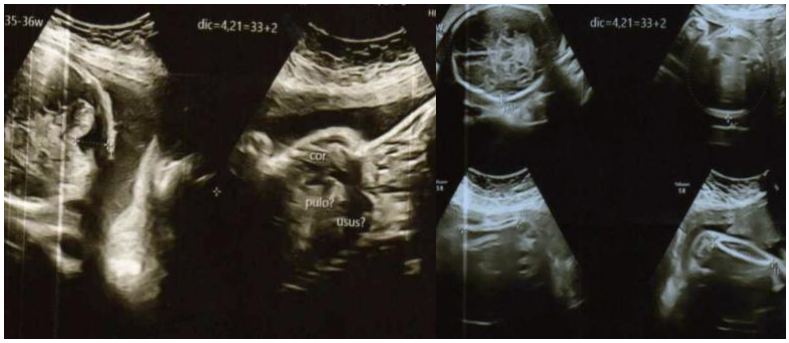

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Figure 2 : The ultrasound showed left sided congenital diaphragmatic hernia

The newborn was born not well and evolved with sudden respiratory distress, requiring ventilation. On auscultation of the left hemithorax, bowel sounds were heard and heart sounds were heard on the right side of the thorax. Chest X ray was done which showed left sided diaphragmatic hernia as shown in (Figure.3) .

The newborn should be examined for murmurs, to be conducted as a congenital and/or abdomen dug present for diaphragmatic hernia, being intubated immediately once he presents any clinical diagnostic. Approximately two hours after birth, the newborn had a sudden respiratory dysfunction evolved into a prolonged cardiac arrest. The newborn was intubated and received cardiac massage and resuscitation. He was then taken to the Intensive Care Unit and carried out a chest radiograph, which showed atelectasis of the right lung and left diaphragmatic hernia. He was not in condition to perform as well as surgical correction of the hernia. He died in the second hours of life. Blood tests were collected during ICU admission, however, it was not ready on time. The

patient died before we perform other examinations such as examination without contrast and Simple acute abdomen X-ray, Echocardiography was. Thus, it was requested the acute abdomen X-ray showed a hemithorax filled cyst like structures (bowel), mediastinum and abdomen relatively free of gas (Figure.3).

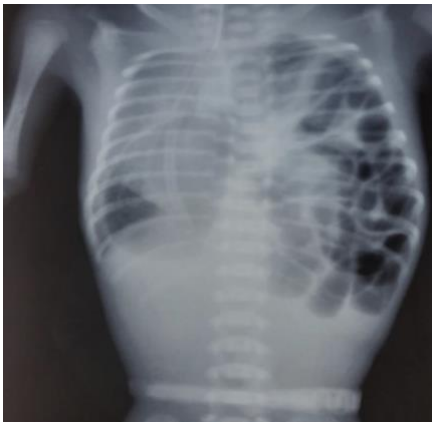

Figure 3. The radiography Abdomen showed left sided diaphragmatic hernia

According to Figure.2, We observe small bowel in the chest through a posterolateral hole, shifting the mediastinum to the right. According to Figure 3. The radiography showed a hemithorax filled cyst like structures (bowel), mediastinum and abdomen relatively free of gas.

DISCUSSION.

Through the diaphragmatic defect of the Bochdaleck hernia, abdominal organs (intestines, spleen, stomach, kidney) may migrate into the pleural cavity, compressing the lung and displacing the mediastinum to the opposite side(1) Depending on the degree of pulmonary compression, there may be a marked decrease of the pulmonary branches, limited multiplication of alveoli, and muscle hypertrophy in pulmonary arterioles, leading to a decreased functional lung mass (pulmonary hypoplasia), involving both lungs with the ipsilateral lung more affected. These newborns are anatomically predisposed to develop Persistent Pulmonary Hypertension, maintaining the permanence of the fetal circulation to the lungs with blood moved through the foramen ovale and ductus arteriosus. The right to left shunt causes acidosis and hypoxia, increased pulmonary vasoconstriction and worsening pulmonary hypertension. It may lead to decreased lung compliance (5).

The clinical profile of respiratory distress reported in our case may progress to severe respiratory distress and lung failure. During physical examination it may be seen excavated abdomen, displaced heart sounds, respiratory failure and bowel sounds in the chest. During the radiography we noted a hemithorax filled cyst like structures (bowel), mediastinum and abdomen relatively free of gas (Figure 3).We should mention the importance of the differential diagnosis with cystic adenomatoid malformation of the lung and pneumonia (6) Infants with CDH often present in the neonatal period with severe respiratory distress, occasionally after a stable period of 24-48 hours followed by acute respiratory distress. Breath sounds are diminished ipsilateral to the hernia. Almost all individuals with CDH have some degree of pulmonary hypoplasia. The

pathogenesis of the pulmonary hypoplasia associated with CDH appears to have both a primary component (i.e., the hypoplasia occurs independent of the diaphragm defect) and a secondary component (i.e., arising from competition for thoracic space particularly in the lung ipsilateral to the hernia). Evidence for the presence of a primary defect in lung development arises mostly from studies in animal models, some of which show that the lung hypoplasia precedes the herniation of abdominal viscera.

Infants with CDH typically require mechanical ventilation and sometimes extracorporeal membrane oxygenation (ECMO) in the newborn period. Major respiratory complications include tracheobronchomalacia, pneumothorax, and secondary lung infection (especially viral pneumonia) that could precipitate terminal respiratory failure even months after surgery. Many infants require ongoing oxygen supplementation and diuretics following surgical correction of CDH. Given the remarkable growth and recuperative capacity of the lung, these treatments can usually be discontinued within the first two years of life.

By early childhood, few children have respiratory symptoms at rest; however, formal testing in older children shows small airway obstruction and diminished blood flow on ventilation-perfusion (V-Q) scan, especially to the lung ipsilateral to the hernia. Reduced exercise tolerance can be a lifelong problem. Intermittent wheezing requiring bronchodilator use is common in people with CDH, and they are at risk for respiratory decompensation with intercurrent illness.

X-ray abdomen and chest standing showed no free gas under diaphragm but right dome of diaphragm was elevated with colonic shadow visible over the liver surface area (Fig. 1). As patient responded well to conservative line of treatment and clinically was not suggestive of peritonitis it was decided to continue the treatment and investigate further.

Treatment in severe cases and prenatal diagnosis is recommended an immediate oxygenation. When it is observed an inadequate prenatal, the morphologic ultrasound examination is the best used during prenatal period. Moreover, once the diaphragmatic hernia diagnostic is done, the newborn should be immediately intubated, in order to perform immediate decompression of the chest with abdominal straps, preventing pulmonary hypertension. All newborns should be immediately intubated after birth or at diagnosis, with the introduction of a nasogastric tube on continuous suction. We should be careful with assisted ventilation in order to maintain a low inspiratory pressure, to avoid damage or breakage of the contralateral lung. Surgical repair should be performed only after stabilization, through subcostal laparotomy with reduction of hernial contents into the abdominal cavity and closure of the diaphragmatic defect. The binomial hypoplasia, pulmonary hypertension remain a major mortality factor (4,7,9,10,11).

CONCLUSION.

Congenital diaphragmatic hernias (CDH) can be diagnosed antenatally by ultrasonography. The diagnosis in the prenatal period induces to a better prognosis, since the birth in a center well equipped will made necessary interventions, minimizing neonatal complications and, thus, optimizing the chances of survival. The clinical importance overlaps the X-ray in case of no diagnosis with prenatal care, it is very important for physicians to be alert for signs that show a possible case of diaphragmatic hernia.

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