J. Dhaka National Med. Coll. Hos. 2014; 20 (02): 61-62

Case report

Congenital Diaphragmatic Hernia

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Abstract :

A 20 hrs. old female baby who was delivered in Dhaka National Medical College Hospital on 9th July 2014 at 12 am was admitted into the neonatal ward with the complaints of severe respiratory distress after birth, Child was restless, irritable and cyanosed. She is the 3rd issue of a non-consanguineous parent. Her other sibs were healthy. She was clinically diagnosed as a case of Congenital Diaphragmatic Hernia and confirmed by chest radiograph. She was given medical treatment as her parents disagree to do surgery.

Introduction :

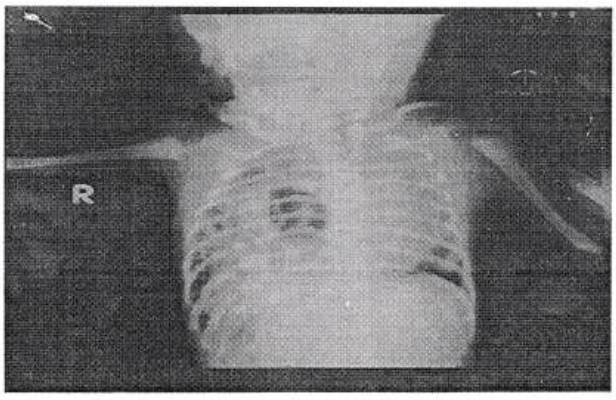
CDH is a congenital malformtion of the diaphragm. The most common type of CDH is Bochdalek hernia, other types include Morgagni Hernia and Hiatus Hernia. The left sided Bochdalek Hernia occurs in 85% cases, left sided hernia allow herniation of both small and large bowel and intrabdominal solid organ into the thoracic cavity.

In Rt sided hernia (13% cases) only the liver and a portion of large bowel tend to herniate. Bilateral hernias are uncommon and usually fatal.[1]

CDH is a life threatening pathology in newborn and a major cause of death due to two complication-pulmonary hypoplasia and pulmonary hypertension[2]. Experts disagree on the relative importance of these two conditions with some focusing on hypoplasia and others on hypertension[3]. Severe respiratory distress and a scaphoid abdomen may be the presenting symptom in a newborn[4].

Case report :

Baby of Shefaly, female baby 20 hrs age born of a non-consanguineous parents admitted into neonatal ward of Dhaka National Medical College & Hospital on 9th July 2014 with the complaints of severe respiratory distress after birth. Baby was irritable, restless and cyanosed. Mother was 35 years old, 3rd gravida, para 3 and had an uneventful antenatal period. The baby was delivered pervaginally at term at Dhaka National Medical College & Hospital. There was no history of membrane rupture nor any other maternal illness, no history of perinatal asphyxia. On anthropometry length 50 cm, weight 3.2 Kg, OFC-35 cm.



On examination the baby was afebrile, heart rate was 150/min. Respiratory rate 100/min. Cyanosis was present. There was no pallor and no icterus, no oedema. There was marked subcostal recession and indrawing of abdomen.

Per abdomen examination revealed scaphoid abdomen; cardiovascular examination revealed that heart sound were better heard at Lt side. Chest auscultation revealed presence of bowel sounds on the Rt side.

Lab investigations :

Hemoglobin 16.6 gm/dl

Blood gas analysis :

pH-7.305

pCO_2-38.4 mmHg, pO_2-49.7 mmHg

HCT-47.3%, Temp 37°C, FIo2-21%, BP-753.8

HCO_3-19.1 mmol/l, O_2 SAT-80%, BE-6.4 mmol/l

S. Electrolyte Na-150 mml/l, K-4.4 mmol/l, ICE -108 mmol/l

Blood group - O^+ ve

X-Ray chest shifting of mediastinum on the left side, free and airfilled loop in the right chest. She was diagnosed as a case of CDH.

Conservative treatment was given; parents disagree to take any medical and surgical Rx. The baby was discharged on risk bond and stayed home and died after 3 days.

Discussion :

Congenital Diaphragmatic hernia occurs in 1 of every 2000-3000 live birth and accounts for 8% of all major congenital anomalies. Most studies report that CDH occurs equally in male and female. The risk of recurrence of isolated CDH in future sibiling is approximately 2% [5].

Seperation of developing thoracic and abdominal cavities is accompanied by closure of posterolateral pluroperitoneal canals. This occurs in the 8th week of gestation. Failure of the canal to close is responsible for these defect.

This condition can often be dignosed before birth and future intervention can sometimes help depending on the severity of condition[6].

A small percentage of cases go unrecognized into adulthood[7].

CDH has a mortality rate 40-62%, outcomes being more favorable in the absence of other congenital abnormalities;

with advances in surgery and care the survival rate is near of 80%[9].

CDH are surgical emergencies; surgery must be performed to remove abdominal organs from the chest and place them back into the abdomen. The diaphragm must be repaired. The ideal time to repair a CDH is unknown. Some authors suggest that repair 24 hours after stabilization is ideal but delays of upto 7-10 days are typically well tolerated and many surgeon now adopt this approach; other surgeon prefer to operate when normal pulmonary artery pressure is maintained for at least 24-48 hours based on echocardiography.

Pharmacotherapy- Use to stabilize BP & circulating volume alleviate pulmonary distress and correct hypoxemia; vasoactive agent (dopamine, dobutamine), opioid analgesic (Fentanyl), Neuromuscular relaxing agents (pancurrnium vecuronium), pulmonary vasodilating agent (Nitric oxide). Medical theraphy in CDH is directed toward optionizing oxygenation while avoiding barotrauma [10]. Placement of avented orogastric tube & connecting it to continuous suction to prevent bowel distension.

Fetal-surgical intervention for CDH may not improve survival compared with standard therapy [11-12]. Postnatal procedures include the following reduction of the herniated viscera and closer of the CDH chest tube drainage in presence of a tension pneumothorax.

In 1946 Gross reported the first successful repair of a CDH in the first 24 hours of life[13]. In the 1960s however Arechon and Reid observed that the high mortality rate of CDH was related to the degree of pulmonary hypoplasia at birth[14]. In recent years evidence suggests that cardiac maldevelopment may further complicate the pathophysiology of CDH[15].

Keller et al found that infants with CDH who have poor outcomes (death or discharge on oxygen) have higher plasma levels of endothelen-1 which is dysregulated in pulmory hypertension[16], the baby will usually be immediately placed on a ventilator. ECMO has been used as part of the treatment strategy at some hospital[17-18].

Sometimes the incision site will be left open to allow the body to adjust to newly moved organs and the pressure associated with that & then closed later once swelling and drainage has decreased [19]. Diaphragmatic eventuation is typically repaired called plication of the draphragm[20].

Conclusion :

Even though CDH is rare, prenatal USG and postnatal chest radiography can diagnose most of causes. Unrecognized diaphragmatic hernia has been the cause of death in infant & toddler. Prompt diagnosis and treatment can prevent serious morbidity and mortality associated with complication.

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