IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 9 Ver. II (September. 2016), PP 156-158
www.iosrjournals.org

Congenital diaphragmatic hernia: a case report and review of literature

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Abstract: *Congenital diaphragmatic hernia (CDH) is an anatomical defect of the diaphragm. It allows abdominal visceras to protrude into the chest and causes serious pulmonary and cardiac complications in the neonatal period. It can be detected antenatally or at birth. The babies usually present with respiratory distress and may also have pulmonary hypoplasia. Timely intervention and proper respiratory management will usually results in good outcome.*
Keywords: *Diaphragmatic hernia; congenital; newborn.*

I. Introduction

CDH is a posterolateral defect of the diaphragm, generally on the left side, through which abdominal viscera herniates into the thorax, displacing the mediastinum to the opposite side.This leads to lung hypoplasia and pulmonary hypertension [1].The incidence of this abnormality is 1 in 2000-5000 live births with a 2% recurrence risk for subsequent pregnancy [2]. Approximately 90% defects occur posterolaterally and 80% on the left [3].The severity and outcomes are determined by the degree of pulmonary hypoplasia and pulmonary hypertension [3]. CDH was described quite earlier but good survival after repair was not achieved until the 20th century. This article describes a case of CDH managed successfully followed by a review of recent literature of this congenital defect.

II. Case report

A newborn male child was referred to our hospital with respiratory distress at birth and decreased breath sounds on the left hemithorax with scaphoid abdomen since birth. Oxygen saturation at presentation was was 85% at room air. There was decreased air entry on left hemithorax with scaphoid abdomen. Bowel sound was heard on auscultation in the left hemithorax. A portable chest radiograph showed mediastinal shift to right and bowel loops in the left hemithorax (Figure 1). Patient was admitted and stabilized in pediatric intensive care unit (ICU) facility and echocardiography was normal. The patient was intubated in ICU and ventilated on high frequency gentle ventilation. At laparotomy a 4 cm defect on the left posterior side of diaphragm. The defect was enlarged and herniated bowel loops were reduced back into the peritoneal cavity. The defect was closed with 4-0 polypropylene sutures.

Postoperatively the patient recovered well and was discharged after 3 days. At 6 months of follow-up, the baby was thriving well.

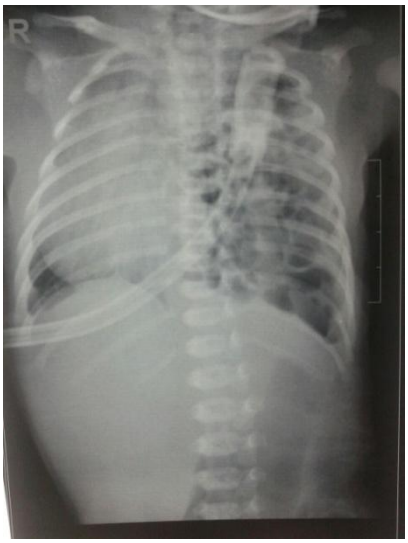

Figure 1 showing bowel loops in left hemithorax

III. Review of literature

While 2% of CDH occur with a familial association, the aetiology remains unclear in most cases. 15% cases occur with some chromosomal anomaly. Recent studies have suggested critical role of chromosome 15q26 for normal diaphragm development [3]. Most cases are isolated;other malformation may occur occasionally [4]. CDH is also observed in non-syndromic chromosomal anomalies like 9p tetrasomy [5]. The delayed or disturbed separation of the thoracic and abdominal compartments of the body leads to orifice in the diaphragm [1]. Ipsilateral lung growth is further impaired by mechanical compression due to herniated abdominal organs; causing interference with respiratory movements and reduction in the secretion of fetal lung fluid [6]. CDH can be detected by ultrasonography during the fetal period which demonstrates presence of abdominal viscera in the thorax. Polyhydramnios may lead to early diagnosis in some severe cases. Further evaluation with lungs- heart ratio [1] and screening for associated congenital anomalies is necessary because associated anomalies confer poor prognosis. Chromosomal anomalies can be ruled out by amniocentesis and lung volume evaluation can be performed by 3-D MRI of the fetus [3]. Clinical features of CDH are respiratory distress with insufficient oxygenation, scaphoid abdomen protruded sternum and mediastinal displacement to the contralateral side [1].Plain X-ray of chest and abdomen shows position of the herniated viscera. Blood gases and pH show the efficiency of gas exchange and helps deriving other indices for better assessment. Echocardiographyhelps to rule out cardiac malformations and measures the right-to-left shunt apart from estimating the severity of pulmonary hypertension. Pulmonary artery diameters and some derived indices may facilitate this task. Echocardiography is one of the more reliable methods for determining when the patient is "stabilized" [1].

Definitive prenatal repair in fetal CDH models has been shown to reverse pulmonary hypoplasia; but forthose cases without liver herniation,it is equivalent to postnatal repair [7]. Another upcoming approach of tracheal occlusion (TO) is based on lung hypoplasia resulting from tracheal atresia. Many observations have been made regarding this after study in fetal models [8]:

1. Increased lung growth in normal and CDH models after TO
2. Increased cell division (with increased alveoli and capillaries) leading to lung growth

Therapeutic tracheal occlusion may be achieved by open tracheal occlusion (small hysterotomy followed by EX-utero Intrapartum Treatment EXIT procedure for securing airway and removing the occlusion clip), endoscopic FEFENDO (endoscopic balloon or clip occlusion of the trachea, also followed by EXIT procedure) and the least invasive FETO (Fetoscopic Endoluminal Tracheal Occlusion: percutaneous method that obviates the need for general anaesthesia and maternal laparotomy) [2].

Postnatal care for CDH requires high frequency gentle ventilation and liquid ventilation with perfluorocarbons if available [2].Nasogastric drainage is required [9] and pulmonary vasodilators like inhaled Nitric Oxide (iNO), PDE5 inhibitors like sildenafil, PGE1, endothelin antagonists and Rho kinase inhibitors help establish adequate perfusion. Extracorporeal Membrane Oxygenators (ECMO) and surfactant provide additional support [2].

Earlier surgical repair of CDH was considered a life-saving emergencybut presently it is undertaken only after cardio-respiratory stabilization. A "delayed" surgery combined with gentleventilation and occasional ECMO support gives thebest results. The duration of delay of surgery is unclear, but few days to weeks maybe beneficial. All preoperative treatments aimto obtain "stabilization" of the patient in terms ofacceptable oxygenation ($PaO_2 > 40$ mmHg) and CO_2excretion ($PaCO_2 < 60$ mmHg) with stable pulmonarypressures (< 50% of systemic pressure), tolerable shunt, good cardiac function and adequate renal clearance on low or no inotrope support. But delayed surgical repair may not always be beneficial [10]. Per-operatively the herniated viscera are carefully reduced into the abdomen and the diaphragmatic orifice is closed with interrupted sutures through a subcostal or transverse abdominal incision.No intercostal tube drainage is necessary. Large defects are closed by a prosthetic patch [11, 12] sutured to the rims of the orifice with interrupted sutures.Patch closure may increase the risk of re-herniation [13-16]. Abdominal wall or latissimus dorsi flaps may be used for CDH repair [17- 20]. Right sided defects and herniation of stomach or liver confer a poor prognosis [2].

Lungs to heart ratio (<0.6 or > 0.6 to 1), low lung volume (25-40%), McGoon Index: right and left pulmonary artery diameters divided by the diameter of descending aorta (cutoff 1.31) and Pulmonary Artery Index: area of right and left pulmonary artery divided by body surface area (cutoff90) are reliable indices for prediction of mortality in CDH [21- 23].

These recent advances have dramatically improved the survival of CDH in the western world but in countries like India, where most nurseries are not equipped with iNO, HFV or ECMO the prognosis is still poor. Scant published data from India pertaining to CDH outcome is another issue that needs to be addressed. For neonates who present within 24 hour of life, survival ranges from 0 to 46% whereas for those who presented after 24 hr was more than 80% [24, 25].

IV. Conclusion

Congenital diaphragmatic hernia may be detected prenatally or at birth. A thoroughphysical examination, correct interpretation of radiological imaging and surgical correction is lifesaving and curative.High degree of suspicion is required for the early diagnosis and proper management.

Conflicts of interest: none

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