REPAIR OF CONGENITAL DIAPHRAGMATIC HERNIA AND EVENTRATION OF DIAPHRAGM WITHOUT VENTILATORY SUPPORT
SAJID MM¹, RAHMAN MAM², CHOWDHURY TK², FAROOQ AA¹, ADNAN WALID³

Abstract:
Background: Congenital Diaphragmatic Hernia (CDH) and eventration of Diaphragm (ED) are important causes of respiratory distress in children. CDH and ED are more a medical emergency than a surgical one and after birth, confirmation of the diagnosis should be followed by treatment in a Neonatal Intensive Care Unit (NICU) or Special Care Neonatal Unit (SCANU), Paediatric Intensive Care Unit (PICU) to stabilize the cardiovascular system. Preoperative resuscitation and delayed surgical repair with or without the use of ExtraCorpororeal Membrane Oxygenation (ECMO) improves the survivability.

Methods: The medical records of all patients with CDH and ED between January 2010 and December 2015 were retrospectively reviewed. Patients’ presentation, management, operative findings and complications were evaluated.

Results: 22 patients were diagnosed as CDH or ED. Male to female ratio was 4.5:1. Age range was 2 days to 7 years, median 82.5 days. Eighteen patients were CDH and 4 patients were ED. Six patients had associated malrotation of the gut. 3 patients had Congenital Heart Disease, 1 patient had Gastrochisis and another had Hiatus hernia. Four patients expired.

Conclusion: Surgery preceded by a preoperative respiratory resuscitation and stabilization reduces postoperative mortality and increases the survival rate. Outcome was not very unsatisfactory without adequate ventilatory support.

Key words: Congenital Diaphragmatic Hernia, Eventration of diaphragm, Ventilatory support, Persistent Pulmonary Hypertension.

Introduction:
Congenital Diaphragmatic Hernia (CDH) is a congenital or developmental defect in the diaphragm through which the abdominal viscera or contents herniate into thoracic cavity. Eventration of Diaphragm (ED) is a pathological elevation of the diaphragm resulting in a paradoxical movement of the affected hemidiaphragm during respiration. The reported incidence of CDH is estimated to be 1 in 2000 to 5000 births. Approximately, one third of babies with CDH is stillborn and is mostly due to fatal congenital anomalies. When stillborns are compared with live births, females are more commonly affected than males. The exact aetiology of CDH is unknown. The recurrent risk in a first-degree relative has an expected rate of 2%. Approximately, 80% of the defects are on the left side and 20% are on the right side. Bilateral CDH are rare and have a high incidence of associated anomalies.

Infants with isolated CDH are more likely to be premature, macrosomic, and male; about one third of affected infants may have associated major defects. Genetic abnormalities associated with CDH include both chromosomal numbers (Turner’s syndrome; trisomies 13, 18, 21, 22, 23) and chromosomal aberrations (15q24-q26 deletions). Several anomalies include lung Hypoplasia, malrotation of gut, cardiac
malformations, and patent ductus arteriosus is present. Although the severity of PH and PPH are major determinant of prognosis and survival, infants with CDH and another major defect have a greater morbidity and mortality. CDH can be diagnosed during fetal life in routine checkup of pregnant women by screening Ultrasonography (USG) which shows guts and liver protruded into the thoracic cavity, polyhydramnios, absent or intrathoracic gastric bubbles, mediastinal shift, and foetal hydrops. The diaphragmatic defect may present as a completely open defect between the abdomen and chest or may be covered by a hernial sac made of peritoneal and pleural membrane. This may mimic evagination of diaphragm which results from birth injury to phrenic nerve or anterior horn cell or phrenic nerve degeneration. This can be excluded by fluoroscopy of the chest where diaphragm moves paradoxically during respiration. Here we present our experience of managing patients with CDH and ED.

Materials and Methods:
This retrospective study was carried out in the department of Paediatric Surgery of Chittagong Medical College and Hospital from January 2010 to December 2015. The collected data was compiled, edited, stratified and analysed for age, sex, presenting symptoms and signs, diagnosis, major associations, operative procedure followed, postoperative complication, post discharge follow up, and necessary comment. The data were collected from previous history records of the department of Paediatric surgery. The patients were referred either from neonatology/paediatrics ward or directly admitted to our ward from emergency/outpatient department. After receiving the patients, they were assessed for vitality, necessary nasal O₂ was given, and resuscitated with intravenous fluid. Required antibiotic was injected. Then, Ultrasonography of abdomen and chest, X-ray of abdomen and chest, ECG, Echocardiography, serum electrolyte, and arterial blood gas (ABG) analysis were carried out. Some patients had high arterial PCO₂ for which NICU were required to stabilize the patient. No ECMO were used because of lack of this facility in this hospital. After, normalizing the blood gas, the patient was prepared for operation. The approach was left or right subcostal incision according to the side of pathology.

Result:
During the 8 year study period 22 patients were diagnosed as CDH or ED. Out of them, 8 were neonates, 10 were infants, and 4 were children. Age range was 2 days to 7 years (median 82.5 days). Eighteen patients were male, and 4 were female. The male: female ratio is 4.5:1. Out of these 22 cases, 18 cases were CDH, of which 16 (88.89%) were left sided diaphragmatic hernia and 2 (11.11%) cases were on right side. The Left: Right ratio was 8:1. Rest 4 cases are ED. The CDH: ED ratio is 4.5:1. Of the 4 ED cases, 3 evolutions were on right side and 1 was on left side. The Left: Right ratio is 1:3.

17 patients had been suffering from respiratory distress, 3 patients had cough, 3 patients had cyanosis, 2 patients had been suffering from pneumonia, 1 patient developed G.I.T. obstruction, 1 patient had vomiting while 1 patient developed hiccup, and 2 patients were reluctant to feeding.

6 patients with CDH had associated malrotation of the gut. 3 patients had Congenital Heart Disease (CHD, 1 atrial septal defect, 1 cyanotic congenital heart disease, 1 dilated atra with dilated ventricle) associated with CDH. 2 of the patients had dextrocardia, and only 1 patient had gastrochisis associated with CDH. 1 patient (age is 4 and half years) underwent laparotomy for Hiatus hernia while incidentally and curiously he was found to have CDH associated with it.

All repairs were achieved via abdominal approach. The small intestine was the most frequently herniated viscerum followed, in order, by the stomach, spleen, colon, and liver. Tension free primary closure by suturing the edges of defect with nonabsorbable 1’0 or 2’0 prolene was reserved for small defects keeping the membrane of sac (if any present) within the sutures of repair. The reconstructive technique using prerenal fascia, costal structures, or different abdominal or thoracic muscle flaps was used in moderately large types of defects.

10 of the patients had the hernial orifice of Bochdalek repaired without any Ladd’s procedure while 4 patients underwent the repair operation with Ladd’s procedure. Of these 14 patients, 6 patients had overlying sacs or membranes which were plicated while repairing the sac. Plication of the diaphragm was carried out in 4 patients with ED. One of these patients had his associated hiatus hernia repaired. Postoperative chest x-ray showed and confirmed correction of deformity and mediastinal shifting with expansion of lung. The CBC and serum electrolyte level was normal. During the postoperative period, 1 patient developed burst abdomen at 10th pod, and another patient developed sepsis at 12th pod. During the post discharge follow up, 17 patients had smooth and uneventful life at 1 and 3 months follow up visit. Only 1 patient developed intestinal obstruction 2 months after operation for which he was admitted in our ward. He was treated conservatively and got cured.
<table>
<thead>
<tr>
<th>Sl. no.</th>
<th>Age, Sex and body weight</th>
<th>Diagnosis at presentation with side affected</th>
<th>Presenting Symptoms and signs</th>
<th>Major Association</th>
<th>Operative procedure</th>
<th>Post operative complication</th>
<th>Follow Up after discharge</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4 months, Male, 4.5 Kg</td>
<td>CDH(left)</td>
<td>Respiratory distress, cough, Reluctant to feeding</td>
<td>Nil</td>
<td>Repair of defect</td>
<td>Nil</td>
<td>Smooth and Uneventful</td>
<td>PICU support not required</td>
</tr>
<tr>
<td>2</td>
<td>8 days, Male, 2.1 Kg</td>
<td>CDH(left) with Malrotation, Term, Low Birth Weight(LBW)</td>
<td>Respiratory distress, excessive crying</td>
<td>CHD(Atrial septal defect)</td>
<td>Repair of defect with Ladd's procedure</td>
<td>Nil</td>
<td>Smooth and Uneventful</td>
<td>NICU support not required</td>
</tr>
<tr>
<td>3</td>
<td>1 month, Male, 4 Kg</td>
<td>CDH(left) with Malrotation</td>
<td>Respiratory distress, Reluctant to feeding</td>
<td>Dextrocardia</td>
<td>Repair of defect with overlying sac with Ladd's procedure</td>
<td>Nil</td>
<td>Do</td>
<td>PICU support not required</td>
</tr>
<tr>
<td>4</td>
<td>10 days, Male, 2 Kg</td>
<td>CDH(left) with Malrotation, Term, LBW, Early Onset Neonatal Sepsis(EONS)</td>
<td>Respiratory distress</td>
<td>Repair of defect with Ladd's procedure</td>
<td>Nil</td>
<td>Do</td>
<td>Patient expired postoperatively at 14 days of age due to multiple congenital anomalies and sepsis. NICU support sought but couldn't be availed</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>6 months, Female, 5 Kg</td>
<td>CDH(left) with Malrotation</td>
<td>Respiratory distress, cough</td>
<td>Repair of defect with Ladd's procedure</td>
<td>Nil</td>
<td>Smooth and Uneventful</td>
<td>PICU support sought but couldn't be availed</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>8 months, Male, 5.5 Kg</td>
<td>ED(right)</td>
<td>Respiratory distress, cough, fever</td>
<td>Plication of diaphragm</td>
<td>Nil</td>
<td>Do</td>
<td>PICU support not required</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>8 days, Female, 2.8 Kg</td>
<td>CDH(left)</td>
<td>Respiratory distress,</td>
<td>Repair of defect</td>
<td>Late onset neonatal sepsis on 12th Pod</td>
<td>Do</td>
<td>NICU support not required</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>12 days, Male, 2.8 Kg</td>
<td>CDH(left)</td>
<td>Respiratory distress,</td>
<td>Repair of defect</td>
<td>Do</td>
<td>NICU support not required</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>16 months, Male, 9 Kg</td>
<td>ED(right)</td>
<td>Respiratory distress, cough,</td>
<td>Plication of diaphragm</td>
<td>Do</td>
<td>PICU support not required</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>3 months, Male, 4.9 Kg</td>
<td>CDH(left)</td>
<td>Severe respiratory distress</td>
<td>Repair of defect with overlying sac</td>
<td>Do</td>
<td>PICU support not required</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>9 days, male, 2.2 Kg</td>
<td>CDH(left) Term, LBW, Postnatal asphyxia (PNA), Hypoxic Ischemic Encephalopathy(HIE-Ill)</td>
<td>PNA, convolution, Lethargy, neonatal jaundice, severe respiratory distress</td>
<td>Operation couldn't be done due to sepsis</td>
<td>Operation couldn't be done due to sepsis</td>
<td>Patient expired at 23 days of age despite vigorous resuscitative effort owing to multiple congenital anomalies</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>2 days, Female, 1.7 Kg</td>
<td>CDH(left) Preterm, LBW, Respiratory distress Syndrome(RDS),</td>
<td>Respiratory distress, Peripheral cyanosis, Sepsis</td>
<td>Gastroachisis</td>
<td>Operation couldn't be done due to sepsis</td>
<td>Patient expired at the age of 3 days owing to sepsis. NICU support sought but couldn't be availed</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 1

Table showing details of patients.
<table>
<thead>
<tr>
<th>Sl. no.</th>
<th>Age, Sex and body</th>
<th>Diagnosis at presentation with side affected</th>
<th>Presenting Symptoms and signs</th>
<th>Major Association</th>
<th>Operative procedure</th>
<th>Post operative complication</th>
<th>Follow Up after discharge</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>5 days, Male, 2.88 Kg</td>
<td>CDH(left) Term, Normal birth weight(NBW), EONS</td>
<td>Respiratory distress, Central cyanosis,</td>
<td>Cyanotic Congenital Heart Disease</td>
<td>Operation couldn’t be done owing to parents’ refusal to operation</td>
<td></td>
<td></td>
<td>DORB</td>
</tr>
<tr>
<td>14</td>
<td>7 days, Male, 2.3 Kg</td>
<td>CDH(left) IUGR, Term, LBW, PNA (III)</td>
<td>Respiratory distress, Peripheral cyanosis, Hiccups</td>
<td></td>
<td>Operation couldn’t be done due to sepsis</td>
<td></td>
<td></td>
<td>Patient expired at age of 14days owing to multiple congenital anomalies NICU support sought but couldn’t be availed</td>
</tr>
<tr>
<td>15</td>
<td>40 days, Female, 3 Kg</td>
<td>CDH(left) with Malrotation of Gut</td>
<td>Respiratory distress, Cyanosis during crying</td>
<td>Dilated right atrium and right ventricle</td>
<td>Repair of defect with Hernial sac</td>
<td></td>
<td></td>
<td>Do</td>
</tr>
<tr>
<td>16</td>
<td>15 months Male, 10 Kg</td>
<td>ED(left)</td>
<td>Recurrent bouts of Pneumonia</td>
<td>Plication of Diaphragm</td>
<td>Smooth and Uneventful</td>
<td></td>
<td></td>
<td>PICU support required</td>
</tr>
<tr>
<td>17</td>
<td>3 months, Male, 5 Kg</td>
<td>CDH(right)</td>
<td>Cough, Fever</td>
<td>Repair of defect with Hernial sac</td>
<td>Smooth and Uneventful</td>
<td></td>
<td></td>
<td>PICU support not required</td>
</tr>
<tr>
<td>18</td>
<td>4.5 years, Male, 9 Kg</td>
<td>ED(right) with Hiatus Hernia</td>
<td>Vomiting, Generalized weakness</td>
<td>Hiatus hernia</td>
<td>Plication of diaphragm with repair of hiatus hernia</td>
<td>Nil</td>
<td>Smooth and Uneventful</td>
<td>PICU support not required</td>
</tr>
<tr>
<td>19</td>
<td>4 months, Male, 6.5 Kg</td>
<td>CDH(left) with Malrotation of Gut</td>
<td>Cough</td>
<td>Repair of defect with Hernial sac</td>
<td></td>
<td></td>
<td></td>
<td>Do</td>
</tr>
<tr>
<td>20</td>
<td>7 years, Male, 15 Kg</td>
<td>CDH(left) with intestinal obstruction( owing to obstruction of transverse colon in the diaphragmatic defect)</td>
<td>Abdominal pain, Vomiting</td>
<td>Restoration of gut from entrapment and Repair of defect</td>
<td>Burst Abdomen on 10th Pod, repaired</td>
<td>Smooth and Uneventful</td>
<td></td>
<td>PICU support not required</td>
</tr>
<tr>
<td>21</td>
<td>6 months, Male, 6 Kg</td>
<td>CDH(right)</td>
<td>Respiratory distress with RTI</td>
<td>Repair of defect with sac</td>
<td>Nil</td>
<td>Smooth and Uneventful</td>
<td></td>
<td>PICU support not required</td>
</tr>
<tr>
<td>22</td>
<td>2.5 months, Male, 3.3 Kg</td>
<td>CDH(left)</td>
<td>Respiratory distress, Cough</td>
<td>Dextrocardia on Echo</td>
<td>Repair of defect</td>
<td>Nil</td>
<td>Do</td>
<td>PICU support not required</td>
</tr>
</tbody>
</table>

Fig.-1: Chest X-ray Postero-anterior and Left lateral view showing bowel gas shadow in left chest.  
Fig.--2: During laparotomy gut is retracted to show the hemial defect of diaphragm before and after repair.
Operative procedure couldn’t be carried out in 4 patients. Out of these 4 patients, 3 patients were suffering from multiple congenital anomalies with sepsis and succumbed to death despite vigorous resuscitative efforts while the attendants of 1 (one) patient refused the operative treatment and left the hospital. ECMO couldn’t be provided because of lack of this facility in our hospital. NICU/PICU supports were not required in 13 patients, either preoperatively or postoperatively; while only 3 patients were provided with NICU/PICU supports with the consultation of anesthetists. Despite severe necessity, NICU/PICU supports couldn’t be availed for 4 patients but those patients survived with manual ventilatory support and intensive care provided by the attendants of the patients.

4 patients expired owing to multiple anomalies and sepsis; out of them, only 1 patient received operative treatment and died postoperatively, other 3 were too ill to receive any treatment.

Discussion:

CDH is mainly a neonatal surgical disorder with multiple congenital anomalies. The earliest description of gross anatomy and pathophysiology associated with CDH was by McCaulay in the proceedings of the Royal College of Physician in 1754. Cooper in 1827 and Laennec in 1834 reported clinical description gross pathology of CDH and suggested laparotomy as the proper approach for reduction and correction of hernia. Bowditch in 1847 first made the bedside diagnosis of CDH and emphasized the clinical criteria for diagnosis. Successful repair of CDH was first reported by Ladd and Gross in 1940 when 9 out 16 patients survived surgery. Genetic abnormalities associated with CDH include both chromosomal numbers (Turner’s syndrome; trisomies 13, 18, 21, 22, 23) and chromosomal aberrations (15q24-q26 deletions). Several anomalies include lung Hypoplasia, malrotation of gut, cardiac malformations, and patent ductus arteriosus is present. Although the severity of PH and PPH are major determinant of prognosis and survival, infants with CDH and another major defect have a greater morbidity and mortality. Hypoplastic left ventricle with hypoplasia of aortic arch is the most common cardiac anomaly which may exacerbate pulmonary hypertension, right to left shunting, and haemodynamic instability. Other cardiac anomalies are atrial septal defect, ventricular septal defect, and transposition of great vessels, tetralogy of Fallot, double-outlet right ventricle, and aortic coarctation.

Neural tube defects are most common central nervous defects. Although, in most of the cases, the patient presents in the neonatal period, the disease may be diagnosed during the 2nd and 3rd trimester by USG or may even present in infancy or childhood. CDH is associated PH and PPH. The other congenital anomalies associated with CDH makes it difficult to manage in one stop surgical station. The morbidity or mortality has not changed much despite the advances made in critical care. In our retrospective study, we will try to compare CDH and ED treated in a tertiary hospital with other studies reported in various publications.
All cases of CDH in this study were approached through an abdominal incision. Reports from CDHSG suggest that the subcostal laparotomy is the most common approach for repair (91%)\textsuperscript{18}.

Of the 18 CDH cases, 16(88.88%) were on left side and 2(11.11%) cases were on right side. The Left: Right ratio is 8: 1. In another study Johannes, Elke and coworkers found 82% were left sided while 18% right sided \textsuperscript{19}. In one study conducted by Clark, Hardin, and co-workers, the ratio is 4:1, which is more or less equal to our observation\textsuperscript{20}. In our study, 18 patients were male, and 4 were female. The male: female ratio is 4.5: 1 which is compatible with another study done by Torf et al\textsuperscript{21}.

Management starts immediately after the patient reaches the hospital. If the diagnosis is confirmed, then immediate preoperative resuscitation includes, opening of an infusion channel, arterial blood gas analysis, and conventional mechanical ventilation (CMV), inhaled nitric oxide administration, and Extra-Corporeal Membrane Oxygenation (ECMO) in severely hypoxic patients with respiratory failure. Surgical repair of CDH can only be considered if arterial blood gas level attains a normal level. In our hospitals with the constraints of Neonatal Intensive Care Units (NICU) facility and non-availability of ECMO, stable patients can be treated by surgery with postoperative mechanical ventilation if needed, which gives a good result. Ours is a study in which we have studied 20 cases, with satisfactory result and follow up. All of them were first surgically repaired and few patients required post-operative NICU facility. In follow up, all of the patients did good result.

In our study we found 17 patients suffering from respiratory distress and 3 patients had cyanosis. Wilson, Bedoyan and coworkers also reported the same symptoms and signs\textsuperscript{22}. In our study, all patients were primarily treated by conventional ventilation with a death rate of 18.18% (4 patients out of 22 patients expired), while in another study done by Ozdogan and Durakbasa, mortality rate had been 50% using conventional ventilation\textsuperscript{23}.

6 patients had associated malrotation of the gut 15(68.18%) patients had isolated CDH and, 7 patients (31.81%) had associated malformation. Of these, 3 patients had Congenital Heart Disease (CHD, 1 Atrial Septal Defect, 1 cyanotic congenital heart disease, 1 dilated atria with dilated ventricle) associated with CDH. 2 of the patients had Dextrocardia, and only 1 patient had Gastrochisis associated with CDH. 1 patient (age is 4 and half years) underwent laparotomy for Hiatus hernia while incidentally and curiously he was found to have CDH associated with it. Stoll, Alembik, and co-workers showed 60.8% associated malformations\textsuperscript{24}. This difference may be due to differences in methodology and surveillance registries.

Closure of abdominal wall was sometimes difficult because of large hernias which could quickly compromise respiratory function, although, no prosthetic materials were used; instead of that, the patient was put into automatic ventilator for assisted mechanical ventilation. Survival rate was difficult to be assessed due to large variations in practicing various management strategies (ventilatory techniques, inclusion criteria for ECMO etc.), timing of surgery, surgical procedures and the increasing number of patients\textsuperscript{25}. Moreover, the presence of associated anomalies, especially congenital heart disease remains an unfavourable outcome of these children\textsuperscript{26}.

The results (survival rate of 77.27% and mortality rate of 18.18%) can be considered far better in a tertiary centre like ours with limited addressability, facility, technical equipment and experience in comparison to other centres in Europe where Georgescu R et al reported a survival rate of 64.29% and postoperative mortality rate of 35.71\%\textsuperscript{27}.

Evntration of the Diaphragm (ED) is a condition in which all or part of the diaphragm is largely composed of fibrous tissue with only a few or no muscle fibres. It is usually congenital but may be acquired\textsuperscript{28}. Complete evntration almost invariably occurs on the left side\textsuperscript{29} is rare on right side, though in our study, most of the cases (¾) are on right side.

**Conclusion:**

Congenital diaphragmatic hernia is amongst the most severe congenital malformations with high mortality caused by its associated pulmonary hypoplasia, persistent pulmonary hypertension and other severe malformations (esp. cardiac anomalies, neural tube defect, and malrotation etc.). Delayed surgery preceded by preoperative respiratory resuscitation and stabilization reduces postoperative mortality and increases the survival rate. The pre and post operative care can also be carried out and patient can be survived without a mechanical ventilatory support in a set up where ICU facility is not available.
References:


