Eventration of Diaphragm – Embryological basis

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Abstract: Eventration of diaphragm refers to the abnormal elevation of dome of the diaphragm. It is a rare anomaly with an incidence of 1 in 10,000 live births. A retrospective study has been done on patients with eventration who presented in Dayanand Medical College & hospital. The signs, symptoms and operative findings have been noted. Recurrent chest infection was found to be the commonest presenting complaint patients. The development of diaphragm with a special reference to the embryological rationale behind eventration has been discussed.

Introduction -
Eventration of the diaphragm refers to an “abnormal elevation of one leaf of an intact diaphragm as a result of paralysis, aplasia or atrophy of varying degrees of muscle fibres (Fig. 1). The unbroken continuity differentiates it from diaphragmatic hernia.” Thomas (1970). It is a rare anomaly, occurring in about 0.001-0.003% of live births, Urschel et al (1989). An eventrated diaphragm may result from congenital etiologies; muscular hypoplasia or aplasia. Acquired causes include phrenic nerve injury during breech delivery, operative trauma (crush, transection and hypothermia), fibrosis, inflammation and neoplasia Stauffer and Rickman (1972). Expedient diagnosis of neonatal diaphragmatic eventration is imperative as the neonate relies upon the diaphragm for 90% of the normal respiratory excursion Connors et al (1991).

In the present paper a retrospective study has been done of patients in paediatric age group, with congenital eventration, who presented in Dayanand Medical College & hospital over a period of 15 years.

Material And Method
Data were collected of all the patients who had been diagnosed and treated for eventration of diaphragm in Dayanand Medical College and hospital over a period of fifteen years. Signs, symptoms, operative findings and associated anomalies were noted. The embryological rationale behind eventration is discussed.

Results
Thirteen cases were studied in all. All the cases were children below 9 years. The youngest being one day old. Respiratory symptoms predominated in patients, (twelve out of thirteen cases presented with either respiratory distress or recurrent chest infection). One of the case presented with abdominal pain due to gastric volvulus, and eventration was a pre-operative finding.

Eleven of the thirteen cases studied were left sided. In two of the cases there was extensive mediastinal shift. Heart sounds were heard on the right side.

Twelve of these cases were repaired surgically. Laprotomy was performed and plication of the elevated dome was done. One of them after symptomatic improvement, refused surgery, went home and was lost to follow up. Postoperative course was uneventful in eleven of the patients. Complications in form of wound infection and incisional hernia occurred in one patient each. Follow up x-rays showed no eventration.

Discussion
The diaphragm develops from a variety of mesenchymal populations between 8th-10th weeks of intrauterine life Sadler (1990). It receives contribution from the Septum transversum, Pleuroperitoneal membranes, Dorsal mesentery of oesophagus and Striated muscle mass, Hamilton & Mossman (1972).

The transverse septum is the primordium of the central tendon of diaphragm. The liver develops in its

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Fig.1: X-Ray (PA View) of the patient showing eventration of diaphragm. Arrow marks elevated dome of diaphragm.
caudal part and its cranial part helps to form the diaphragm. After the formation of head folds during the 4th week, the septum transversum forms a thick incomplete partition between the pericardial and abdominal cavities. Posterior to the septum transversum, however, the pleural and peritoneal cavities communicate through the pleuroperitoneal canals that lie on either side of oesophagus. The partition between the thorax and abdomen is completed when the pleuroperitoneal canals are closed by the formation of pleuroperitoneal membranes.

Pleuroperitoneal Membranes fuse with the dorsal mesentery of oesophagus & septum transversum. This completes the partition between the thoracic and abdominal cavities. Although the pleuroperitoneal membranes form large portions of the foetal diaphragm, they represent relatively small portions of the newborn infants diaphragm, Moore & Persaud (1999).

The dorsal mesentery provides a central point of fusion for the septum transversum and the pleuroperitoneal membranes. It forms the median portion of diaphragm. The crura of the diaphragm develop from myoblasts that grow into the dorsal mesentery of the oesophagus. During fifth week, myoblasts from third to fifth cervical somites migrate into developing diaphragm bringing their nerve fibres (phrenic nerve) with them. At weeks nine to twelve of gestation, the pleural cavities expand upto lateral body wall and finally penetrate into the wall. This process has been called burrowing by Bremer (1943). This splits the wall of the body into two layers outer and inner. Outer layer which forms thoracic wall and inner layer which becomes incorporated into diaphragm. Accordingly, muscular material from thoracic myotomes also enters diaphragm. This configuration explains that the peripheral part of diaphragm receives innervation from lower six intercostals nerves.

Two types of embryologic failure may be responsible for eventration, failure of myoblasts to migrate between the two leaves of the pleuroperitoneal membrane on the affected side and non innervation of myoblasts.

Eventration therefore results from non muscularisation or non innervation or both Sadler (1990), Moore and Persaud (1999). Usually unilateral it is commoner on left side.

In present study also, congenital eventration was probably due to muscular hypoplasia; in varying degrees, from a thin muscle plate to its complete absence, leaving only a thin membrane consisting of pleura, connective tissue and peritoneum.eleven out of thirteen cases were left sided.

Other congenital malformations like pulmonary hypoplasia or aplasia, cardiac defects, hydrocephalus etc.may be associated with congenital eventration. In our study, two of the patients presented with extensive mediastinal shift, one had gastric volvulus and rest had no associated congenital anomalies.

Neonates poorly tolerate paradoxical hemidiaphragmatic motion and the resultant mediastinal flutter. By comparison, older children and adults may remain asymptomatic with unilateral eventration and no ipsilateral diaphragmatic function, Connors et al

Table: I Clinical Profile Of Patients With Eventration Of Diaphragm

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Presenting Symptoms</th>
<th>Associated Abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>7.5 months</td>
<td>M</td>
<td>Respiratory Distress</td>
<td>None</td>
</tr>
<tr>
<td>2.</td>
<td>8 months</td>
<td>F</td>
<td>Recurrent Chest Infections</td>
<td>None</td>
</tr>
<tr>
<td>3.</td>
<td>3 months</td>
<td>M</td>
<td>Respiratory Distress</td>
<td>None</td>
</tr>
<tr>
<td>4.</td>
<td>6.5 months</td>
<td>F</td>
<td>Respiratory Distress</td>
<td>None</td>
</tr>
<tr>
<td>5.</td>
<td>9.5 months</td>
<td>M</td>
<td>Recurrent Chest Infections</td>
<td>None</td>
</tr>
<tr>
<td>6.</td>
<td>11 months</td>
<td>M</td>
<td>Recurrent Chest Infections</td>
<td>None</td>
</tr>
<tr>
<td>7.</td>
<td>8 months</td>
<td>M</td>
<td>Recurrent Chest Infections</td>
<td>None</td>
</tr>
<tr>
<td>8.</td>
<td>2 years</td>
<td>M</td>
<td>Abdominal pain &amp; vomiting</td>
<td>None</td>
</tr>
<tr>
<td>9.</td>
<td>9 years</td>
<td>M</td>
<td>Recurrent Chest Infections</td>
<td>None</td>
</tr>
<tr>
<td>10.</td>
<td>1 day</td>
<td>F</td>
<td>Respiratory Distress since Birth</td>
<td>None</td>
</tr>
<tr>
<td>11.</td>
<td>8 months</td>
<td>M</td>
<td>Breathlessness since birth, multiple episodes of vomiting’ 4days</td>
<td>None</td>
</tr>
<tr>
<td>12.</td>
<td>3 days</td>
<td>F</td>
<td>Respiratory Distress since Birth</td>
<td>Mediastinal Shift, Heart sounds heard on Right side.</td>
</tr>
<tr>
<td>13.</td>
<td>8 days</td>
<td>M</td>
<td>Respiratory Distress since Birth, cyanosis/ refused feed’ 1day</td>
<td>Mediastinal Shift, Heart sounds heard on Right side.</td>
</tr>
</tbody>
</table>
Common presentations in patients with significant eventration include: Recurrent chest infections, Severe respiratory distress with cyanosis, Intestinal obstruction following gastric volvulus, Palpitations due to mediastinal shift.

In present study also, respiratory symptoms predominated. Twelve of the thirteen cases presented with respiratory distress.

Eventration can be diagnosed by elevation of the affected dome of diaphragm on chest X-ray (Fig. 1). Other imaging modalities may be required to differentiate it from a hernia. Previously, pneumoperitoneography served as the ‘gold standard’ but has been replaced by fluoroscopy or high resolution ultrasound, Kaplan et al (1994).

Asymptomatic patients are managed conservatively but all patients with significant symptoms require surgery. Paradoxical diaphragmatic movements suggest complete paralysis or severe hypoplasia and is a strong indication of surgery.

The diaphragm is approached via thoracotomy or laprotomy. Thoracotomy is preferred on the right side owing to the presence of liver. Laprotomy is more useful on the left side as associated intra-abdominal conditions like gastric volvulus may be dealt through the same incision. The surgical procedure involves: Plication of the diaphragm, the goal of plication is to restore the diaphragm to the position of midexpiration. If possible, repair is performed using interrupted, nonabsorbable sutures or Excision of the redundant part of the diaphragm and repair. Kimura et al (1991), have recommended use of decron felt pledgets to prevent sutures from cutting through in cases where the diaphragm is too thin to safely hold the suture.

**Reference**


**Fig. 2:** Contrast Xray (both Lateral and PA view) of an eight day old male showing eventration. Arrow marks eventration.