# A STUDY ON 114 CASES OF CONGENITAL DIAPHRAGMATIC EVENTRATION

## DISSERTATION

## Submitted for

## M.Ch (PAEDIATRIC SURGERY) - Branch V Examination

## 2007

**INSTITUTE OF CHILD HEALTH AND HOSPITAL FOR CHILDREN**
**MADRAS MEDICAL COLLEGE**

**THE TAMILNADU DR.M.G.R.MEDICAL UNIVERSITY**

**CHENNAI**

## CERTIFICATE

This is to certify that the dissertation entitled **"A STUDY ON 114 CASES OF CONGENITAL DIAPHRAGMATIC EVENTRATION"** presented here is a original work done by Dr. **C. Aravindan,** M.Ch. Post Graduate in Institute of Child Health and Hospital for Children, Madras Medical College, Egmore, Chennai – 600 008, in partial fulfillment of the University rules and Regulations for award of M.Ch. Degree – Paediatric Surgery, under my guidance and supervision during the academic period from 2004 – 2007.

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## ACKNOWLEDGMENT

I am grateful to our respected Dean of Madras Medical College for allowing me to do this study.

It is my privilege to express my gratitude to **Prof. Kulandai Kasthuri M.D., D.C.H.,** the Director ICH & HC for allowing me to conduct the study at our Institute. I wish to convey my sincere thanks to the Head of our departmenat, **Prof. Jeyakumar M.S., M.Ch.,** for his guidance and support to complete this dissertation.

I am grateful to Prof. Philip Chandran, **M.S., M.Ch.,** for his guidance and advice to properly complete this study.

My sincere thanks are due to Prof. S.V.Senthil Nathan, **M.S., M.Ch.,** for his advice and encouragement to finish this study well.

I am greatly indebted to **Prof. K. Seeniraj M.S., M.Ch.,** who with his vast knowledge, experience, valuable guidance and constant support at every stage of this study has contributed enormously for making this study possible.

I am greatly thankful to **Prof. Rajkishore Bagdi, M.S., M.Ch.,** for his valuable guidance.

I wish to express my sincere thanks to **Prof. G. Iango, M.S. Mch,** for his guidance for this dissertation.

I sincerely thank **Prof. R. Bhaskar** for his advice and support for completing this work.

I am grateful to all the assistant professors in the Department of Paediatric Surgery for their help and support throughout my course and during the study.

I would like to express my gratitude to the great masters – the children of this hospital who have been constant source of knowledge for all who work for them and I dedicate this study for all children who are the cause of all our learning.

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## INTRODUCTION

Diaphragmatic eventration is an abnormal elevation of the intact diaphragm. .It may be congenital or acquired. It accounts for 5% of all diaphragmatic anomalies(1). Congenital diaphragmatic eventration(CDE) by definition present at birth. As with Bochhdalek's congenital diaphragmatic hernia (CDH), congenital diaphragmatic eventration is thought to result from a congenital anomaly of pleuroperitoneal membrane but occurring at a slightly later stage.

The condition remains important from the time it was recognized because of its close clinical similarity with CDH which has been a subject of great interest to pediatric surgeons throughout the world. CDH has seen over the years tremendous improvement in its survival and constant addition of diagnostic and therapeutic armamentarium in its management has made this possible though the cases with severe pulmonary hypoplasia and persistent pulmonary hypertension have very poor survival rates even in this modern era. 15- 20% of all CDH cases have a sac covering the herniated contents and this makes the differentiation between a complete eventration and CDH very difficult. And many

authors say such a differentiation is not essential especially in massive herniation because the prognosis in these cases is

similar because of associated pulmonary hypoplasia(2) and such a differentiation is only arbitrary(3).

The current study concentrates on clinical features, diagnosis, management and prognosis of this relatively rare clinical entity called congenital diaphragmatic eventration and comparison of prognosis of newborn cases of CDH and CDE in Indian scenario, in an institute like ours where we deal with a large volume of cases from all over South India which we thought could be of immense help for anyone who wants to know the clinical behavior of these cases and its management. There are only limited available studies in the literature worldwide with a large series as with this study which includes 114 cases of CDE. This study also aims at reviewing the complication rates of the two commonly performed surgical procedures for CDE individually which is all the more very difficult find in the literature.

**ESSENTIAL ANATOMY OF DIAPHRAGM:**

The diaphragm is composed of a central tendinous area from which muscle fibers radiate in all directions towards their peripheral attachment.

The fan shaped muscle of diaphragm arises from the internal circumference of

the thorax, with attachments to the sternum, the lower six or seven ribs and vertebral bodies of the lumbar vertebrae. Posteriorly the muscle fibers originate from the aponeurotic arch of of the ligamentum arcuatum externum which overrides the psoas and quadratus lumborum muscles. Laterally the fibers of the diaphragm inter digitate with slips of transverse abdominisas they originate from the ribs. The right crus is larger and longer than the left and arises from the bodies of the upper three or four vertebral bodies. The left crus arises from the upper two lumbar vertebral bodies. All the muscular fibers insert into the trefoil shaped cental tendon and this tendon is partially fused with the undersurface of the pericardium.

There are three natural openings within the diaphragm. The aortic opening is the most posterior of all and is formed from fibers comprising the right and left diaphragmatic crura. This tunnel is actually behind the diaphragm and not within it and it contains aorta, the azygos vein and the thoracic duct. The oesophageal hiatus is slightly ventral from aortic hiatus and consists of fibers passing between the aorta and oesophagus towards the right crus. And fibers converging on the pericardial tendon. The opening of the IVC lies within the confluence of the tendons of the right hemithorax and tendon beneath the pericardium.

The pleura is tightly adherent to the top surface of the central tendon and most of the musculature. It is impossible to separate the pleura from the central tendon of each hemidiaphragm .

The superior phrenic arteries are located on the thoracic side of the diaphragm.

They are two in number and branches from thoracic aorta. The other arteries supplying superior diaphragmatic surface are paired pericardio phrenic and musculophrenic arteries which are branches from internal mammary arteries on either side. The major blood supply is from the inferior aspect by inferior phrenic arteries– branches from aorta or celiac axis and arise at the level of median arcuate ligament of the diaphragm. The venous drainage of the diaphragm generally closely follows the arterial supply.

The following observations have been made from anatomic and ultrasonagraphic studies of newborn diaphragm:

1. The diaphragm inserts only on the anterior costo diaphragmatic rib cage border.

2. From anterolaterally to posteriorly the diaphragmatic insertion has increasingly greater distance from the rib cage.

3. The dorsal diaphragm ends its free course at the eleventh rib and continues caudally ending between the twelfth rib and the crista iliaca.

Thus the diaphragm in a neonate acts as bellows moving mainly in the posterior part whereas in adult it acts as piston. The diaphragm of the newborn which has a flat curve because of its large angle of insertion on the rib cage and a small area of apposition can only suck in the rib cage rather than the air. It is this rib cage action which results in increase in chest volume. (5)

## DIAPHRAGMATIC INNERVATION

The phrenic nerve originates from C3, C4and C5 nerve roots and then enter the thoracic cavity in front of the subclavian artery. Majority of the diaphragmatic muscle originates from cervical myotomes innervated by fibers from the spinal nerve roots at cervical levels C3, C4 and C5. These fibers join and form the phrenic nerve which elongates as the septum transversum migrates caudally. There is however an outer rim of diaphragmatic muscle which originates from the migrating mesenchymal cells of the nearby body wall innervted by spinal nerves from T7 to T12.

Despite the contributions from the thoracic spinal nerve roots majority of the diaphragm is innervated by the phrenic nerve. After its mediastinal course the phrenic nerve usually divides at the level of the diaphragm or above it. The right phrenic nerve enters the diaphragm just lateral to the IVC within the central tendon. The left nerve enters lateral to the left border of heart just anterior to the central tendon in the muscle itself. The infra diaphragmatic course can be predicted even if not seen directly by knowing the distributions of the four main motor divisions.

The phrenic nerve first splits into anterior and posterior trunks. The anterior trunk further divides into sternal and anterolateral branches near the anteromedial border of the central tendon. The posterior trunk likewise divides into crural and posterolateral branches near the posteromedial margin of the cental tendon. The sternal and crural branches are short and run medially.The anterolateral and posterolateral branches are longer and run along the muscular insertion margin of the central tendon. These branches innervate majority of the diaphragm. Their anatomical relation to one another is described as a pair of hand cuffs or manacles. These branches within muscle layer and not readily visible.(6)

**EMBRYOLOGY:**

The embryology of the diaphragm remains incompletely understood and involves multiple, complex cellular and tissue interactions. The fully developed diaphragm is derived from four distinct components:

(1) The anterior central tendon forms from the septum transversum,

(2) The dorsolateral portions from the pleuroperitoneal membranes,

(3) The dorsal crura evolve from the esophageal mesentery, and

(4) The muscular portion of the diaphragm develops from the thoracic intercostals muscle groups.

The precursors of diaphragmatic structure begin to form during the fourth week of

gestation with the appearance of the peritoneal fold from the lateral mesenchymal tissue. At the same time, the septum transversum forms from the inferior portion of the pericardial cavity. The septum transversum serves to separate the thoracic from the abdominal cavities and eventually forms the central tendinous area of the fully developed diaphragm. It defines the rudimentary pleuroperitoneal canals and allows for the establishment of mesenchymal tissue within these canals that ultimately results in pulmonary parenchymal development.

Closure of the pleuroperitoneal canals with the formation of a pleuroperitoneal membrane occurs during the eight week of gestation. Several theories have been put forward to explain the formation of this membrane and the subsequent development of a diaphragmatic structure. Progressive growth of the pleuroperitoneal membrane has been one mechanism proposed for canal closure. Other researchers have postulated that concurrent hepatic and adrenal organogenesis is crucial to this process. The involvement of a posthepatic mesenchymal plate in diaphragmatic formation has been proposed. (4)

## CLINIAL FEATURES AND MANAGEMENT OF DIAPHRAGMATIC EVENTRATION AND LITERATURE REVIEW

Congenital diaphragmatic eventration(CDE) is a rare diaphragmatic abnormality caused in utero incomplete muscularization of the membranous diaphragm. The first recorded description goes back to 1774 when Jean Louis Petit noted it on a post mortem examination.(7). It accounts formation 5% of diaphragmatic anomalies.

Congenital diaphragmatic eventration of diaphragm by definition present at birth. It is associated with a lack of normal muscularization of the fused pleuropericardial membrane which forms the diaphragm but it occurs at a later stage in utero than congenital diaphragmatic hernia(8). Whether this phenomenon arises secondary to defective distribution of the phrenic nerve fibers or abnormal muscularization of the primitive diaphragm is not proven.

The most acceptable theory suggests lack of migaration of myoblasts along the phrenic nerve branches as the primary etiology. Eventration of diaphragm can involve a portion of diaphragm or the entire hemidiaphragm or very rarely the entire diaphragm . The phrenic nerve and the site of attachment of diaphragmatic margins are normal. The muscle fibers in the diaphragm are sparsely distributed and nonfunctional, but not

atrophic. As a result the diaphragm is lax, thin, and elongated rising as a smooth arched membrane into the involved hemithorax. Congenital eventration of diaphragm differs from congenital diaphragmatic hernia in that the diaphragm is intact and no communication exists between the thoracic and abdominal cavities.

Acquired or paralytic eventration of the diaphragm can result from a wide variety of causes. (35) According to some series incidence of secondary diaphragmatic paralysis is even as high as 50% (9) These include,

1. birth trauma
2. infection(polio, Fetal rubella , CMV infection)
3. malignant tumour infiltration of phrenic nerve
4. congenital absence of anterior horn cells- Werdnig Hoffman syndrome.
5. operative injury during cervical. Cardiac or mediastinal procedures.

In children birth trauma is by far the commonest cause of acquired paralytic eventration. This is often related to a difficult vaginal delivery with shoulder dystocia or breech birth, leading to a stretch injury to the C3-C5 roots and affecting the origin of phrenic nerve and brachial plexus(Erb-Duchenne or Klumpke' palsy). Fractures of the humerus or clavicle are often seen with these neural injuries and should be sought. A wide spectrum of injury severity is possible. When the nerve roots are avulsed, no

improvement can be expected. Lesser degrees of injury or edema are recoverable with time.

The diaphragm in patients with acquired phrenic nerve paralysis is initially normal with appropriate muscle development and position. With time muscle atrophy develops and the diaphragm becomes attenuated and rises into the hemithorax.

Associated anomalies are commonly seen with pulmonary and gastrointestinal systems in the form of pulmonary hypoplasia and intestinal malrotation respectively. Other anomalies rarely reported include megacolon, hypospadias, situs inversus, congenital heart disease, Ehler Danlos syndrome, cleft palate, tracheomalacia, bony abnormalities and myopathies and chromosomal abnormalities (trisomy 13, 15,and 18) (11,12).The presence of these anomalies are strong argument for congenital etiology of eventration. Diaphragmatic eventrations are most commonly unilateral with only few bilateral cases reported ever in the literature.(19,11). The involvement of diaphragm can be either partial or complete. One of the largest series published by Wayne et al in 1974, 65% of the cases were partial(segmental)eventrations with most of these located on the right, whereas those with complete eventrations were located on the left side.(12) Males are more commonly affected than females varying from1:1 to 4:1(11,13)

**PATHOPHYSIOLOGY AND SYMPTOMS:**

The pathophysiological changes accompanying eventration may involve both the respiratory system and the GI tract.(10). When present symptoms arise as a result of

inadequate ventilation or abnormal migration of abdominal viscera into the evntration. Respiratory problems include lung collapse, mediastinal shift affecting contralateral lung resulting in tachypnoea, distress, tachycardia and cyanosis(13). These symotoms are more severe in the neonates and infants in whom respiratory symptoms increase during feeding and with abdominal distension. A non functional diaphragm is comparable to a flail chest injury from a functional point of view. The pulmonary and haemodynamic effects vary greatly with age of the patient and comorbidities if present. A paralyzed hemidiaphragm whether due to a congenital or an acquired cause, is far more clinically relevant to a newborn baby than to older children or adults. This results from the fact that infants are much more dependant on diaphragmatic excursions than adults for tidal volume exchange. Additionally newborns are subject to the phenomenon of paradoxical respiration, whereby the normal lung is compressed by mediastinum during inspiration. Paradoxical breathing is explained by the fact that the negative pressure generated by the normal side is not counter balanced by the abnormal side especially in newborns who have weak intercostals muscles, soft thoracic cage and a mobile mediastinum. Therefore newborns with a paralyzed or an immobile eventrated diaphragm may develop an acute respiratory failure requiring endotracheal intubation and mechanical ventilation. When occurring post operatively the manifestation of phrenic nerve palsy will be inability to be weaned from mechanical ventilation.

Gastro intestinal problems include abdominal pain, dysphagia, heartburn, belching, etc and intestinal obstruction due to associated malrotation. Some children are

also prone for acute gastric volvulus of organo axial type due to stretched gastrosplenic and gastrophrenic ligaments. It has been reported that 40 to 60% of all pediatric patients with gastric volvulus have diaphragmatic abnormalities of which 20% is eventration. (14)

**DIAGNOSIS:**

Clinical features range from being asymptomatic to severe respiratory distress. Patients may present with repeated attacks of pneumonitis later in childhood.

Dullness of involved hemithorax, decreases breath sounds and features of pneumonitis are hallmarks in physical examination. In most cases eventration is first recognized in a chest X ray taken for respiratory distress which demonstrate characteristic elevation of the involved hemidiaphragm atleast 2 inter costal spaces higher than the other side.(16) Demonstration of paralysis or paradoxical motion of diaphragm requires chest fluoroscopy or ultrasonography. Radio nuclide ventilation perfusion scans often show a 50- 75% decrease in ventilation on the involved side. However this quantification of ventilatory defect not always correlate well severity of symptoms. Radiographic differentiation between CDE and CDH with sac is difficult if

not impossible in extensive cases.

The charecteristic physical and radiological findings are not present in mechanically ventilated children. Positive pressure ventilation reverses the diaphragmatic displacement and pulmonary ventilation appears normal. Chest X rays with the patient breathing spontaneously are necessary to identify the characteristic radiographic finding and should be done in all patients who can not tolerate discontinuation from ventilation.

In addition eventration should be considered in all patients with unexplained respiratory distress and require urgent intubation before baseline chest X ray can be obtained.

**INDICATIONS FOR OPERATION:**

Indications include

1. progressive respiratory distress and atelectasis or Pneumonia
2. eventration associated with birth injury not improving during observation
3. patients who have known phrenic nerve injury during thoracic or mediastinal operations
4. significant elevation in a CDE compromising the growth of ipsilateral lung though there is no significant respiratory distress

Infants who have progressive respiratory distress due to CDE should undergo

surgery without delay as their mediastinal anatomy limits tolerance of this defect. They do not benefit from conservative treatment(15,20). Infants who have birth injury in which there is a potential for recovery exists present a dilemma. Some authors recommend a waiting period of 1 month to allow recovery.(17)

**OPERATIVE CORRECTION:**

**Goals of surgery:**

1. To restore the diaphragm to the normal location within the involved hemithorax.
2. To restore normal capacity to the involved hemithorax to allow normal lung growth in infants.
3. To restore normal visceral location in the abdomen
4. To stabilize the mediastinum by eliminating paradoxical motion of the diaphragm

These goals have been achieved by two different surgical procedures with one using placation and other with resection of the redundant diaphragm. Either procedure should effectively eliminate the passive redundant diaphragmatic tissue and establish in its place a rigid diaphragmatic structure 2 intercostal spaces lower than where it had been. (18)

Diaphragmatic plication was first suggested as treatment of eventration of diaphragm in 1916. (21) However it was only performed first in 1954 by Birmingham(22).

**OPEN PLICATIONAL REPAIR:**

The traditional operative approach uses a posterolateral thoracotomy in most cases. All right sided cases are best approached through chest. Left sided lesions are approached via thoracotomy or laparotomy. Some authors prefer thoractomy because of better visualization of the phrenic nerve branches. Bilateral eventrations though rare are better approached by laparotomy. Patients who present with gastric volvulus or intestinal obstruction should undergo laparotomy .

Many authors prefer simple plication than resection.(12,15,20,23,24). After exposure of the diaphragm the course of phrenic nerve branches should be examinesd if possible. Then two rows of imbricting sutures are placed on the redundant diaphragm. The initial row of plicating sutures are placed along the postero lateral one third of the diaphragm beginning at the central portion of the diaphragm and proceeding towards the periphery. Sutures are placed parallel to the phrenic nerve branches. Each suture placed should pick up several centimeters of tissue in two or three equally spaced bites. During imbrication, diaphragmatic should be lifted off liver surface in case of right eventration of diaphragm.The entire row of interrupted sutures are placed and then tied to plicate the diaphragm .Permanent sutures like 2-0 nylon are better. A second row sutures are then placed from centre along the anterior one third of diaphragm. After plication the

diaphragm should be taut,not redundant. If this has not been achieved the tissue along the suture lines can be plicated again over itself to continue imbrication of the hemi diaphragm until proper correction is achieved. Technical variants include use of stapling devices.(25)

Dr. David State in 1949 described a subcostal radial placation technique for eventration of diaphragm.(26) The original technique included a generous incision across the right upper quadrant of the abdomen and placement of radial sutures along the muscular portion of diaphragm pulling it towards the lateral chest wall. John Foker at University of Minnesota used transthoracic radial placation technique since 1976 to treat 35 children.(27) His technique included interrupted mattress pledgeted sutures imbricating muscular diaphragm via posterolateral thoracotomy in a radial manner towards chest wall extending from xiphoid process upto vertebral body excluding mediastinal pleura.In this series 31 of the 36 surgeries(86%) led to extubation within three days eventhough 15 patients had been ventilator dependant preoperatively. There were no deaths within 30 days. Only one had (3%) recurrence long term and there were 72% long term survival. This in comparison with Smith C series(15%) which shows 19% recurrence for conventional central tendon plication technique has very good results.

A thoracostomy tube is placed for pleural drainage and can usually be removed after 24- 48 hours. Antibiotic coverage is given until chest tube is removed. The patient can be often weaned from ventilator immediately after operation.

## OPEN DIAPHRAGMTIC RESECTION:

Alternative procedure involves resection of redundant portion of the diaphragm. A full thickness ellipse of central tendon of the diaphragm is removed and the remaining portions are imbricated to achieve two layer closure. The diaphragm resected should not include major phrenic nerve branches. The first row of sutures is placed and then tied to overlap the diaphragmatic margins and a second row secures the overlapping margins. Second row of sutures should not transgress diaphragm to avoid abdominal organ injury if the approach is thoracotomy.

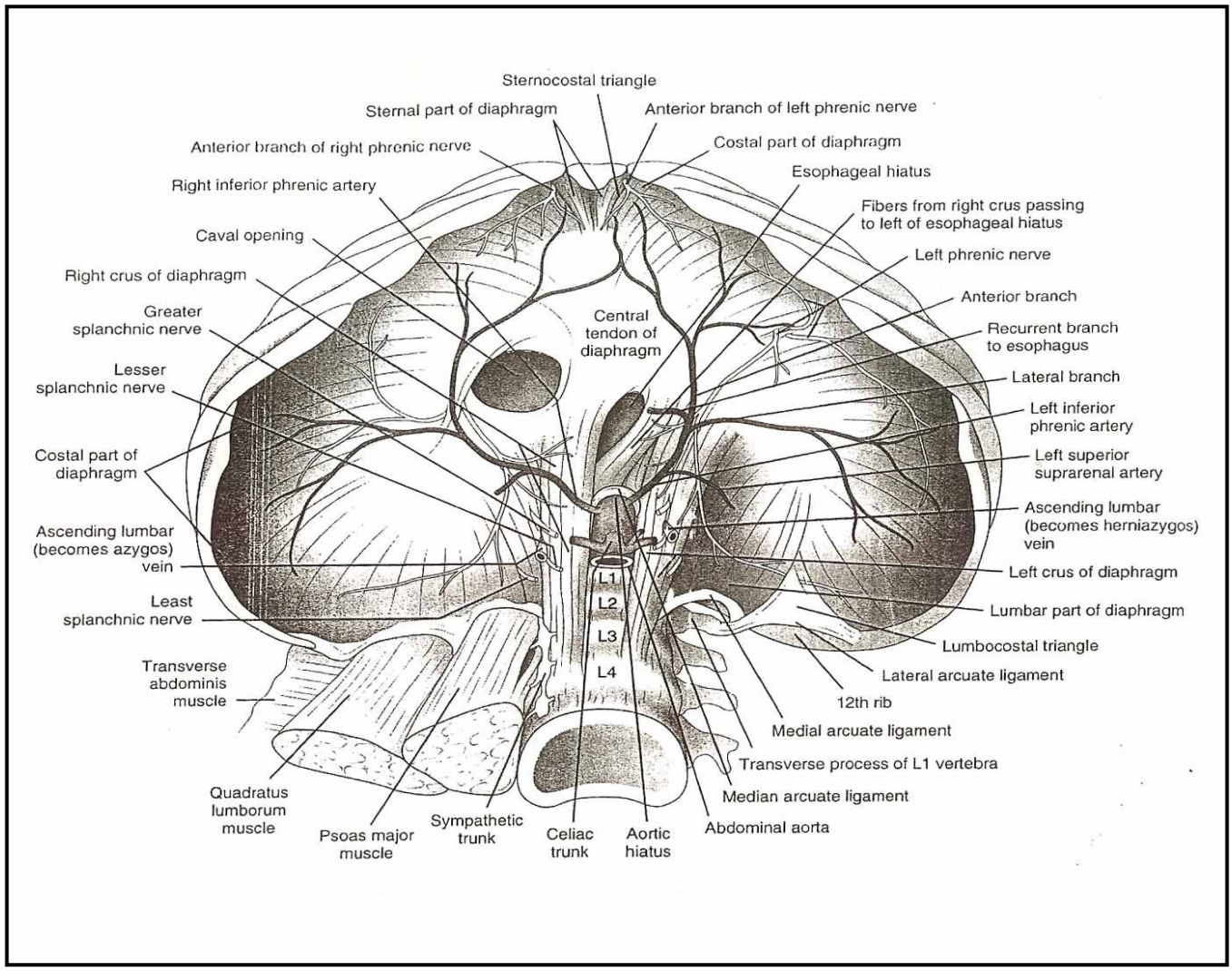
## MINIMALLY INVASIVE REPAIR:

Minimally invasive technique offers a safe and effective alternative formation diaphragmatic placation. Selective tracheobronchial intubation improves visualization excluding ventilation on the operated side. The 0 degree telescope is introduced in the 5th inter costal space at posterior axillary line. Working ports are placed at mid axillary and mid clavicular line along the fifth to seventh inter costal space. If a mini thoracotomy video assisted thoracoscopy approach is used to facilitate suturing this is placed in the posterior axillary line in the ninth ICS and the midclavicular port is excluded. Apex of the diaphragm is grasped with a grasper and displaced into the abdomen and imbricating sutures are placed along the margin of the depressed area starting from periphery towards the centre. Both approaches allow redundancy to be removed as with conventional open plication(28). Minimally invasive radial plication has been used with good results at some centres.(29)

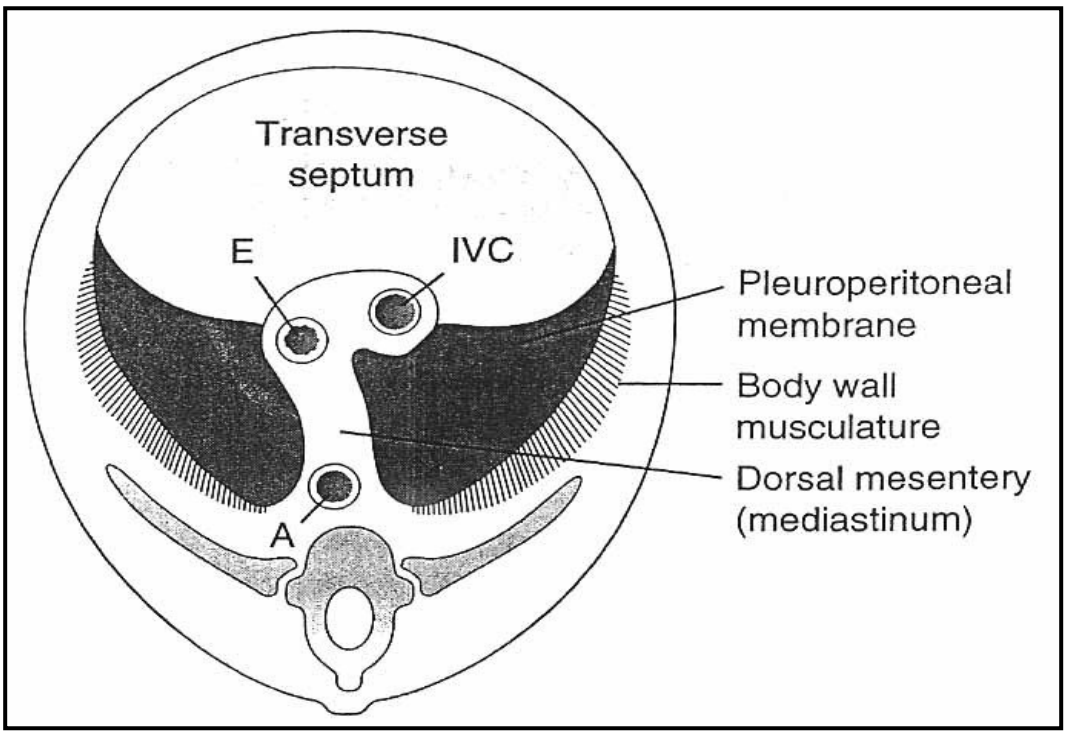
**RESULTS OF SURGERY AND PROGNOSIS:**

In most series the results diaphragmatic repair are dramatic with the patients being extubated within a few days after surgery or even in the operating room.(33).However carefully selected patients is key for success as with any other surgical procedure. Diaphragmatic plication will not prevent future functioning of diaphragm should there be a recovery of nerve transmission in phrenic nerve injury cases.

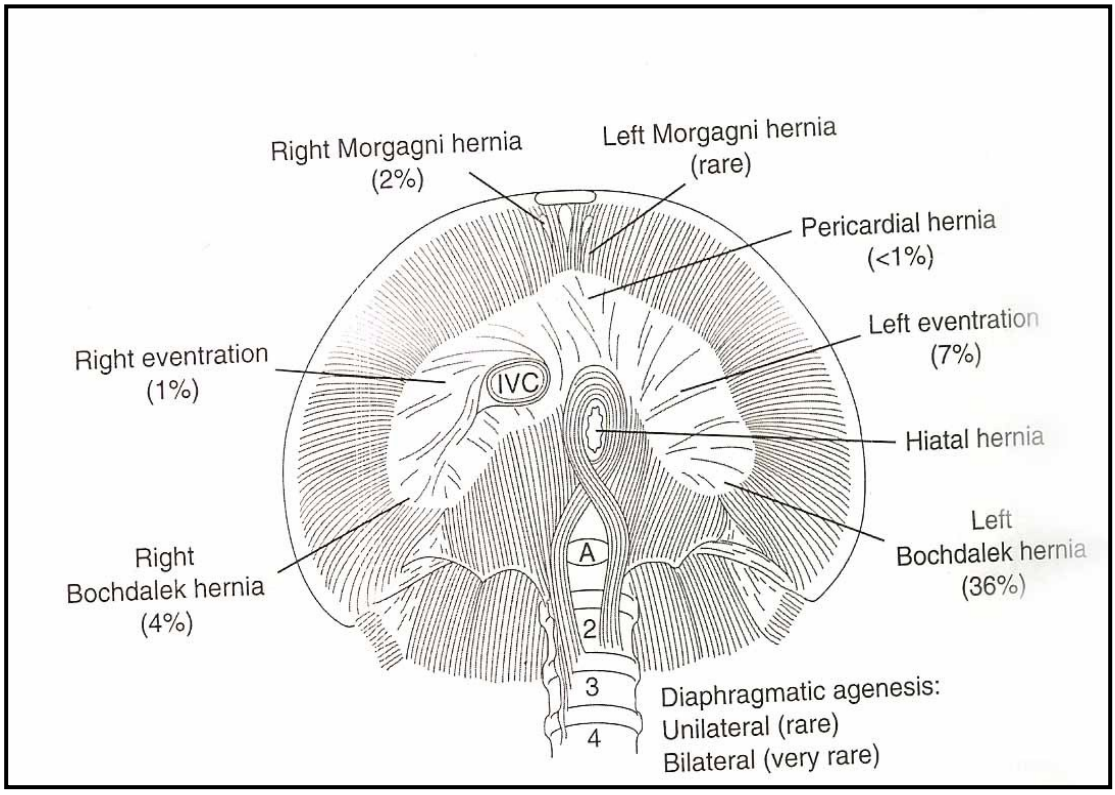
A high mortality rate 14% has been noticed in a large series by Tsugawa et al(32) who advocated early surgery for all cases possibly because of an early surgery done even formation children with serious cardiac anomalies. Other series by Yazicsi et al(33 patients), Tiryaki et al(15 patients), Jaclitsch et al(36 patients), SmithC et al show similar good results with a mortality ranging from 5to 20% for CDE cases and a recurrence rate for plicational repair ranging from 3% to 19%.(15,27, 30, 31,32) Specific complications rates for resectional repair has not been given in any of these studies nor the specific long term effects of diaphragmatic surgeries like GERD, chronic lung disease etc.


Sternocostal triangle
Sternal part of diaphragm
Anterior branch of left phrenic nerve
Anterior branch of right phrenic nerve
Costal part of diaphragm
Esophageal hiatus
Right inferior phrenic artery
Fibers from right crus passing to left of esophageal hiatus
Caval opening
Left phrenic nerve
Right crus of diaphragm
Anterior branch
Greater splanchnic nerve
Central tendon of diaphragm
Recurrent branch to esophagus
Lesser splanchnic nerve
Lateral branch
Left inferior phrenic artery
Costal part of diaphragm
Left superior suprarenal artery
Ascending lumbar (becomes herniazygos) vein
Ascending lumbar (becomes azygos) vein
L1
L2
L3
L4
Left crus of diaphragm
Least splanchnic nerve
Lumbar part of diaphragm
Lumbocostal triangle
Transverse abdominis muscle
Lateral arcuate ligament
12th rib
Medial arcuate ligament
Transverse process of L1 vertebra
Median arcuate ligament
Quadratus lumborum muscle
Psoas major muscle
Sympathetic trunk
Celiac trunk
Aortic hiatus
Abdominal aorta

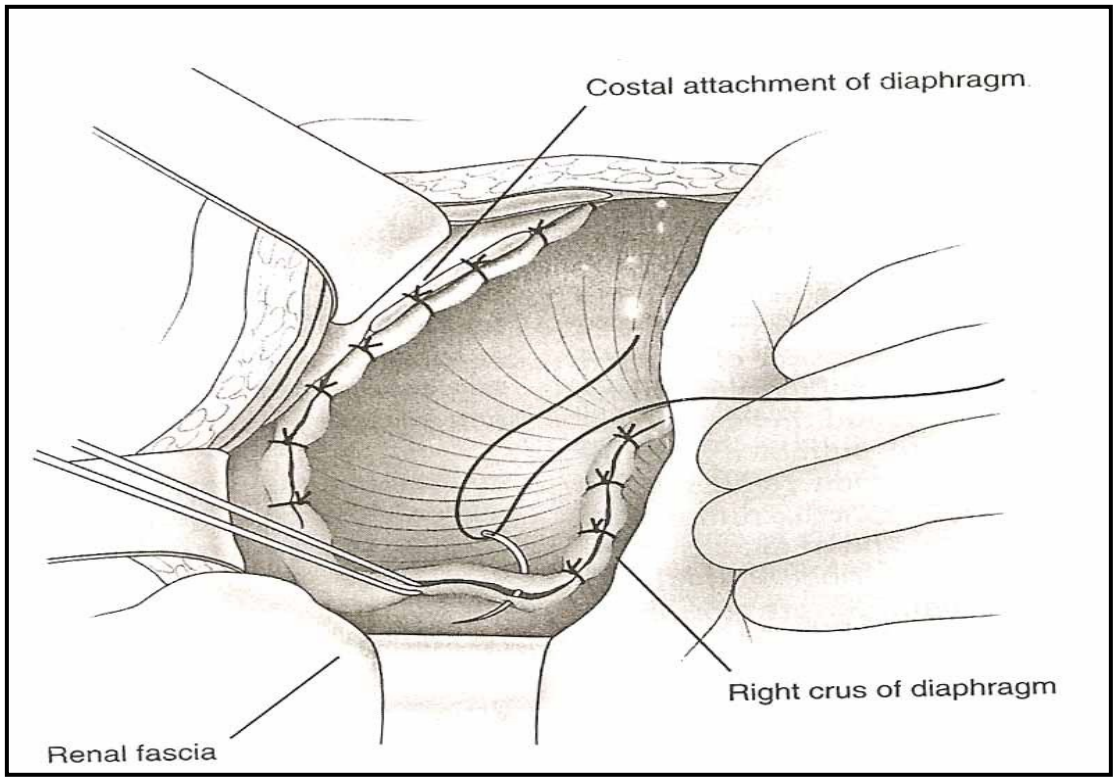
## Embryological Components of the Diaphragm



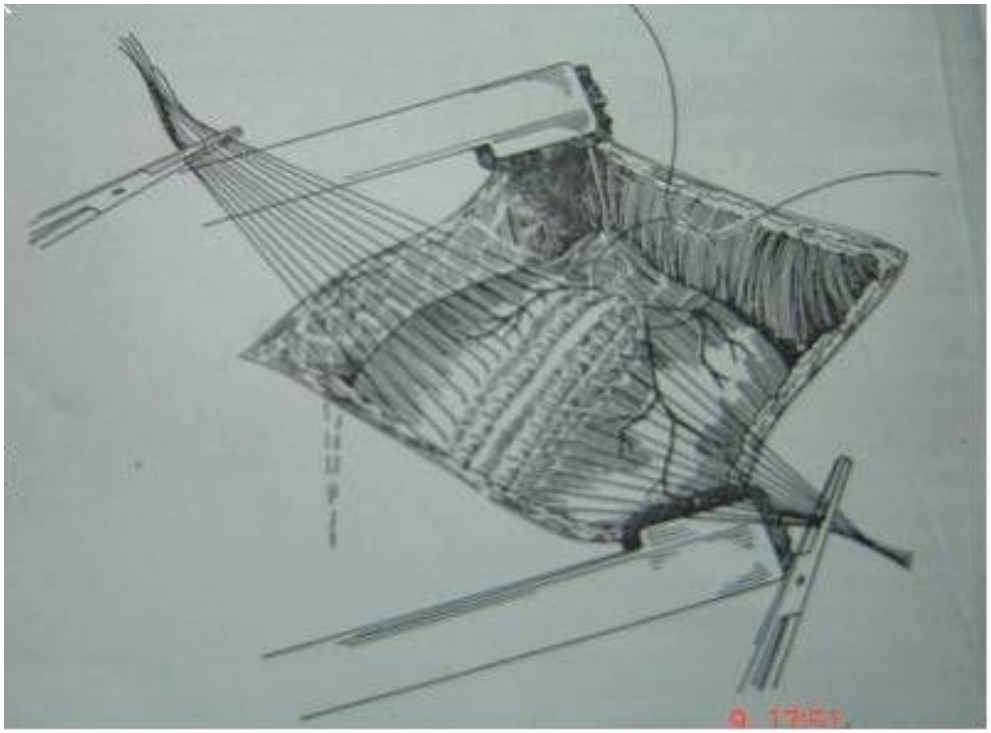

## Distribution of Congenital Lesions of the Diaphragm



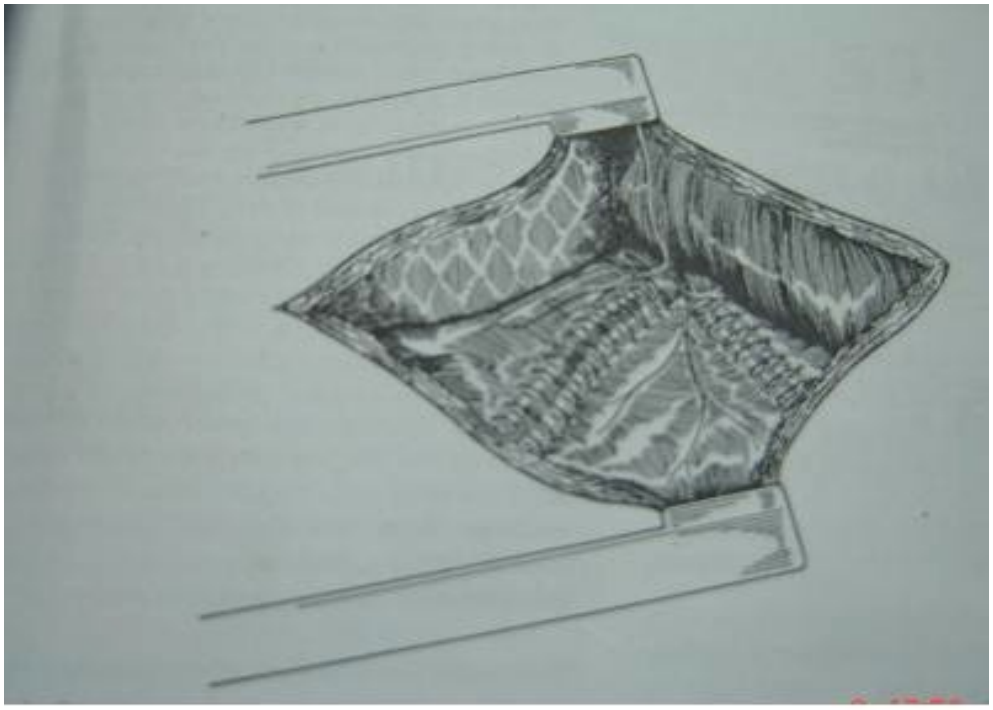

**State's Original Radial Plicational Technique**



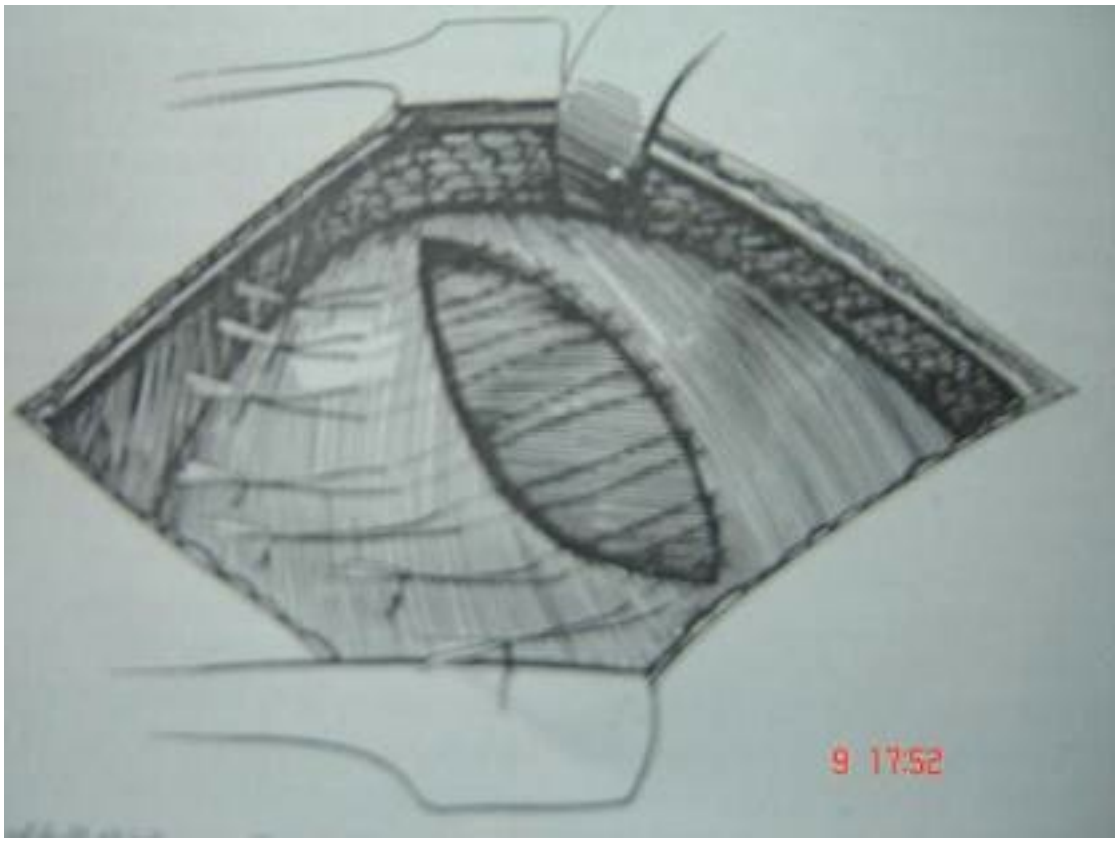
**Diaphragmatic Plicational Repair with sutures in position**



**Diaphragmatic Plicational Repair – Completed**



# Diaphragmtic Resection and Repair




## AIM OF THE STUDY

1. To review the various clinical presentations of cases of diaphragmatic eventration and to know the common modes of presentation in various age groups

2. To study the surgeries performed for diaphragmatic eventration and their results and to have a literature review in the subject

3. To evaluate the prognosis following surgery and to study the prognosis of

congenital diaphragmatic eventration in comparison with congenital diaphragmatic hernia in the newborn period

## MATERIALS AND METHODS

All cases of diaphragmatic eventration diagnosed and treated at our hospital during the period from March 2001 to March 2006 (6 year period) were studied by reviewing the case records retrospectively. There were totally 114 cases.

Age of presentation, sex, chief complaints and clinical features were noted. Chest X ray findings based on which the preoperative diagnosis was made were recorded. Note was made on other investigations done. Operative procedures done and the intra

operative findings were noted. Complications and follow up of the cases were also recorded.

In older children the diagnosis was fairly straight forward based on chest X ray. In neonates and infants with massive herniation of abdominal contents the clues conventionally used in  to distinguish eventration from diaphragmatic hernia in our institute were coiling of NG tube inside the thoracic cavity (which is a strong point in favour of eventration) and presence of a well defined margin above the contents suggesting a sac covering it along with a clinical status which is some what better in cases of diaphragmatic eventration.

Further distinction was made intra operatively by an absence of well defined rim of diaphragm with a defect which is placed more posteriorly . The excised portion of diaphragm during repair was sent routinely for HPE in an attempt to classify the cases as CDE and CDH.

Comparison clear cut cases of diaphragmatic eventration presented in the neonatal period was done with cases of CDH in terms of preoperative and postoperative mortality and long term morbidity.

## ANALYSIS AND DISCUSSION

### INCIDENCE:

Of the 114 cases 40(35%) presented in the neonatal period, 52(45%) presented within 3 months of age and nearly 79% of the cases presented within 3 yrs of life. With recent extensive usage of antenatal screening for congenital anomalies 6 cases had been detected antenatally; but they were all diagnosed as CDH as in both CDH and CDE mediastinal shift can be seen as well as presence of abdominal viscera in the thorax. A confident diagnosis of CDE can be made only if a leaflet of diaphragm can be made and that is very difficult with ultrasound examination. MRI appears to be a promising tool

for this purpose.(37, 38)

CDE was found to be more common among males in all age groups. The sex incidence was male: female 1.5:1(male 68 and female 46) Left sided eventration is more common than the right sided cases. There were 87 left sided cases and 27 right sided cases falling to a ratio of L:R- 3:1. 27(23.7%) cases had only segmental involvement even on one side and 17 were right sided in that. There were no bilateral cases in this series.

**ETIOLOGY:**

All the cases in this series are classified as congenital diaphragmatic eventarations as there were no discernible causes which would lead to secondary diaphragmatic palsy. There was no obvious history of birth trauma in any of these cases. Wherever excision and repair of diaphragm was done the intactness of the phrenic nerve was confirmed where possible.

**CLINICAL FEATURES:**

About 50% of the children older than 3months presented with respiratory infections and eventration was identified during routine CXR. And in this group 80% of the cases had previous frequent respiratory infections. Of the remaining 50% (31 children) 22 were asymptomatic and eventration was found during routine CXR for

single episode of respiratory infection or for unrelated conditions like foreign body ingestion or abdominal xray for an unrelated abdominal colic.

One area of interest is the unusual presentations like gastric volvulus in the eventrated portion of diaphragm (4cases), intestinal obstruction due to malrotation (2cases), failure to thrive and associated GERD in 4 cases. The cases with gastric volvulus presented with non projectile vomiting following feeding and upper abdominal distension and VGP. There was difficulty to

pass NG tube in 2 cases and NG tube was passed freely in another 2.In these 2 cases because of easy passage of NG tube gastric volvulus was not considered initially but later volvulus was confirmed during surgery and the stomachs in these children were partly gangrenous requiring gastrostomy and one of these children died ultimately of sepsis.

Though malrotation was a frequent finding in many cases of complete eventration during surgery it was not typically a cause for presentation in many cases. Only 2 cases presented with intestinal obstruction due to Ladd's bands and prompt operative repair of diaphragm and correction of malrotation was done in both the cases with good results. Eventration was picked up during evaluation for failure to thrive and GERD symptoms in 4 cases.

In neonates and children less than 3 months three types of presentation were

typically noticed.

1. Those presenting with respiratory distress and tachypnoea almost indistinguishable from CDH within hours after birth(30 cases)- most of them referred from where the babies were delivered which includes the antenatally diagnosed cases. The mortality in this set was high- 16%

2. Those presenting later in the newborn period with respiratory distress and admitted in medical wards as mucous plug syndrome, pneumonitis or sepsis and most of them requiring ventilatory support (8 cases). These cases had difficulty in weaning from ventilator in spite of adequate management of underlying conditions and there was prompt improvement in cases which underwent operative repair after recognizing eventrations .Significantly in one of these children eventration could not be made out in CXR because of positive pressure due to mechanical ventilation and recognized later when the child was slightly better and was off the ventilator. However the mortality in this group was very high 50%.

3. Third group is the “late presentors”(14 cases)in new born with minimal respiratory distress and no complications and having excellent prognosis of all and nil mortality. This group includes 2 cases of segmental eventration

   Total mortality in neonates is 10 cases ie 19% mortality in this age group.

**ASSOCIATED ANOMALIES:**

Our children were not extensively evaluated for associated anomalies as the literature is not supportive. Cardiac evaluation was done after surgery in neonates with precordial murmurs. Older children had no associated anomalies in this series.

Neonates had 11.5%(6 cases) associated cardiac anomalies. Another 5 neonates were not evaluated because of very bad general condition and they died even before surgery. Cardiac anomalies included VSD in 3cases, PDA in 1 and ASD in 2. Two cases had associated proximal hypospadias and one case had an associated cleft palate. No other anomalies were found either in the perioperative period or in the follow up.

Incidence of congenital anomalies and involved systems are almost similar to the current study in another major series by Sarihan H et al(39). In his series an association with pulmonary segmentation abnormalities have been noticed and such an association is associated with very grave prognosis.

**INVESTIGATIONS:**

Chest X ray was done in all cases and in most of the cases that was the only investigation done. It has been policy of the institute to take X rays with nasogastric tube in place to know the location of stomach and presence of coiled NG tube in chest

suggesting presence of stomach inside the chest is strongly in favour of eventration. This point has been proved by the fact that out of 32 cases of newborn complete left sided CDE 26 had stomach in the thorax during surgery. Whereas in a review of our 50 cases of CDH stomach was found only in 4 cases .

Fluoroscopy was used only in 12 cases to look for paradoxical movement and that too in older children. And use of that additional investigation did not alter the management.Similarly ultra sound was used in 16 cases to look for paradoxical movement. These two investigations which are said to be useful in neonatal cases to differentiate from CDH could not be done regularly in our set up. In the neonatal cases the preoperative diagnosis of CDE was confirmed during surgery in 35 out of 47 cases(74.4%). The combination of good clinical condition of the baby, definite upper margin of the herniated contents suggestive of a covering by stretched diaphragm and coiled NG tube in thorax suggesting stomach located there is strongly in favour of CDE than CDH.

**MANAGEMENT:**

Though many techniques have been described in literature for diaphragmatic repair in CDE in our institute the surgeries for CDE were mainly plication or partial excision and repair. All cases diagnosed as CDE were operated except a few children. These cases included 5 neonates who died before surgery and 8 older children ( 6 segmental cases and 2 complete varieties with mild elevation) and 2 of them were not operated because they were not willing for surgery but 4 of them subsequently operated

due frequent respiratory infections and other 4 cases (all segmental varieties) are still being followed up(mean follow up of 2.1years).

23 right sided cases were operated. All right sided cases were operated by thoracic approach by $6^{th}$ or 7th intercostal space except 4 which were approached by right subcostal laparotomy. Reason for preferring laparotomy in these cases is not known. All left sided cases were operated by left subcostal laparotomy.

Of the 82 left sided cases simple plication was done in 61 cases and partial excision and repair in remaining 21 cases. Similarly of the 23 operated right sided cases placation was done in 14 cases and excisional repair was done in 9. The reason for choosing a particular procedure for an individual case is not known.

The cases which presented with gastric volvulus underwent laparotomy by left subcostal approach and derotation and no specific measures to fix stomach other than diaphragmatic repair. 2 cases had gastric necrosis and gastrostomy was done and one of them died of sepsis post operatively.

Malrotation which was a common association of most cases of complete left sided eventration was dealt with by dividing Ladd's bands if any and appendectomy.

The excised portion of the diaphragm was sent for histopathological examination in most of the cases. In a total of 21 such HPE examinations diapharagmatic muscle fibers were present in 16 of the cases and mere fibrous tissue and connective tissues in rest of the specimens. This includes 6 older children in whom the diagnosis of CDE was

almost unmistakable during surgery with a central deficiency and stretched diaphragm. This suggests that a differentiation of CDH and CDE based on HPE findings to distinguish a sac structure and stretched muscle is not possible. And clinical relevance the differentiation between a congenital diaphragmatic hernia with a sac and CDE is not clear.

**MANAGEMENT TABLE OF ALL CASES OF CDE:**

The above chart depicts the management done in the set 114 cases of CDE. 9 out of 114 cases were not operated forming about 7.8%. In the ‘less than 3 months age group’ surgery was done in 5 children because of poor general condition they ultimately succumbed. In the older age group surgery rate was almost 93.5%. The remaining 6.5% were managed conservatively.

TYPES OF DIAPHRAGMTIC REPAIR AND SIDE DISTRIBUTION:

The following chart depicts the types of surgeries performed formation CDE in our institute. Simple placation was done in 71.4% cases. 60.8% of all right sided cases

and 74.3% of left sided cases underwent plicational repair. Apparently the use of this repair has been more in left sided cases.

| | **SIMPLE PLICATION** | **PARTIAL EXCISION AND REPAIR** |
|---|---|---|
| **RIGHT SIDED** | **14** | **9** |
| **LEFT SIDED** | **61** | **21** |
| **TOTAL** | **75** | **30** |

-

**PROGNOSIS**:

Prognosis appears good in older children. Whereas in neonates the mortality was 19% (10 cases) half of which died even before surgery because of a sick general condition at presentation. Overall mortality was 10.5%(12 cases) and 8.7% of which were children less than 3 months of age.

The two older children who died were a 2yr old male child and 3.5 yr old male child; the former died after a long illness following gastrostomy done for gangrenous

stomach due to volvulus and the latter had adhesive intestinal obstruction 3mths after initial CDE surgery and laparotomy and adhesiolysis was done , developed intestinal fistula and died of sepsis.

Another large series by Tsugawa etal, in which all the 50 children were operated including the17 childen who presented with respiratory failure and preoperative ventilation. Long term follow up in the 43 patients who survived showed diaphragmatic position indistinguishable from normal children. Two patients required replication and one child died 3 months after surgery. Five of the 25 patients who had phrenic nerve injury died: two from cardiac anomalies, two from congenital motor neuron disease and one from pulmonary failure. Of the 25 patients who had congenital muscular deficiency. Two died from cardiac complications

**COMPLICATIONS:**

Complication rate in this series was 21.9% and most of them were minor.

Complications are as follows,

1. Wound infection - 14(13.3%)

   (includes wound dehiscence)

2. Partial wound dehiscence - 6(5.7%)

   (requiring suturing)

3. Adhesive intestinal obstruction - 8(7.6%)

Operated - 4

(one died of fistula and sepsis)

Conservative - 4

4. Recurrence and reoperation - 3(2.8%)

5. Pneumothorax (ipsilateral- left) - 3(2.8%)

6. Empyema - 3(2.8%)

Wound complications were the commonest and 6 cases had partial wound dehiscence requiring secondary skin suturing. Adhesive intestinal obstruction following CDE surgery seems to be quiet high (7.6%) and half of them requiring laparotomy and adhesiolysis. Significantly 6 of the eight cases who had adhesive obstruction had undergone excisional repair. It requires critical analysis because there are several studies which insist simple placation in all cases of CDE(Wayne et al, Smith et al, Kizilcan et al,Haller et al).

Recurrence in the form of elevated diaphragm more than 2 inter costal spaces than the opposite side had to be operated in 3 cases. All these cases had undergone plicational repair. It has been policy of the institute to slightly over correct during plicational repair in the recent years to avoid this complication. In Tsugawa et al series the recurrence rate for placation was 4%. In Smith C et al study the reelevation rate for open plication was as high as 19%.(15) In John Foker's series of 36 patients who underwent trans thoracic radial plication technique 31 were extubated within 3 days though 15 patients were ventilator dependant preoperatively. There were no deaths within 30 days of surgery and no morbidity directly related to plication. 26 of these patients survived long term(12 years median follow up) and diaphragmatic function was reevaluated in 18 patients using ultrasound and some degree of function had returned to 14 cases.

Most of the cases had a chest tube following surgery for CDE and 14 left sided cases did not have a chest tube following plication and one of them developed pneumothorax on the same side which was promptly recognized by routine post operative chest X ray. Whereas the other 2 cases of pneumothorax occurred on contralateral side and they were also promptly managed by chest tube insertion.

3 cases developed empyema postoperatively 2 in excisional cases and one in plicational case. Cause in these cases is unknown but chest tube could have been the cause of empyema.

**COMPARISON OF PROGNOSIS OF NEONATAL CASES OF CDE AND CDH:**

Though it is generally believed that newborn eventrations behave clinically similar to that of CDH our experience in this series has been that CDE neonatal cases which require ventilatory support before surgery improve after surgery unlike CDH cases which require prolonged post operative ventilatory support and with high mortality.

In our series of 96 cases of CDH in the same 5 years 47 were antenatally diagnosed and overall mortality rate was 64% whereas the mortality in this series of CDE is 19% in the newborn period. Whether this can be taken as an evidence to say that lung development is not grossly abnormal as with CDH in CDE with resultant

pulmonary hypoplasia and persistent pulmonary hypertension require further evaluation. Further evaluation very important in our set up is because we do not routinely use echocardiogram, frequent ABGs or ECMO.

In our series the prognosis and survival of CDE appears to be good and is almost similar to another major series by Garbaccio et al.(36)

## SUMMARY

114 cases of CDE diagnosed and treated over period of 5 years from March, 2001 to March 2006 were retrospectively studied. The results are as follows:

Of these cases 40(35%) presented in the newborn period. 52(45%)presented within 3 months of life and almost 79% of cases presented before 3 years of life. There were 6 antenatally diagnosed cases, but they were diagnosed as CDH.

Male to female ratio in this series was 1.5:1. Left to right ratio was 3:1.About 23.7% cases were segmentally involved predominantly involving right side.

There were no discernible primary lesions causing secondary diaphragmatic palsy

in this series.

About 50% of children older than 3 months presented with recurrent respiratory infection and in another 50% the diagnosis was made incidentally on CXRs.

Unusual presentations like gastric volvulus(4 cases) and intestinal obstruction due to malrotation(2 cases) were noticed.

In the neonatal period the presentation is almost similar to CDH. In this group the children were typically falling into three groups viz. early presentation similar to CDH(30cases), sick late presentation(8 cases) and uncomplicated late presentation (14cases). Prognosis was best with the third group.

Associated cardiac anomalies were noted in 11.5% of the newborn cases.

Chest X ray is the first and only investigation in most of the cases. Presence of stomach in the chest, delineation of a clear sac margin in the chest X ray and a good clinical condition of the baby put together strongly favour the diagnosis of CDE than CDH. Using these clues a confident diagnosis of CDE was preoperatively in 74.4% of the cases.

105 cases underwent surgical repair. 5 newborn children died before surgery and

2 older children are being followed up without surgery. Right sided cases were operated by thoracotomy through 6th or 7th inter costal space and left sided cases were approached by left subcostal approach.

Major complication rate in this series is 21.9% which includes wound dehiscence(5.7%), adhesive intestinal obstruction requiring surgery(3.8%), recurrence(2.8%), pneumothorax 92.8%), empyaema thoracis(2.8%). Chest tue was kept in 86.7 % of the cases. Adhesive intestinal obstruction seems to be commoner with excisional repair and recurrent eventration more in plicational repair cases.

The mortality appears to be much lower with CDE(19%) than with CDH(64%).

## CONCLUSIONS

1. 35% of all CDE cases present in newborn period and 79% present before 3 years of life.
2. Left to right ratio is 3:1 and male to female ratio is 1.5:1. Bilateral cases are very rare.
3. Segmental eventration was found in 23.7% cases and it usually affects right hemidiaphragm.
4. Diaphragmatic elevation secondary to a primary lesion or due to birth injury

is very rare.

5. Cardiac anomalies are the commonest association – 11.5% in this series.

6. CXR is the most important investigation required for diagnosis and is sufficient in most of the cases. A confident diagnosis of CDE was made in 74.4% of the cases according to this series with a combination of CXR and clinical features.

7. Simple plication of diaphragm(71.5%) was the commonest surgery for CDE followed by excisional repair(28.5%) in our hospital.

8. Major complications rate is 21.5 %. Wound complications are the commonest. Adhesive intestinal obstruction is commoner in excisional repair cases and recurrence of eventration appears to be common with placation. It needs further evaluation before calling it as a significant association.

9. Mortality rate according to this series is 10.5% and the rate is almost double in the newborn period.

10. Prognosis of newborn CDE cases as compared to CDH cases is much better probably due to less commoner association of lung hypoplasia with CDE cases.

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**PROFORMA**

Name : Age : Sex :

DOA : DOD : IP :

(i) Detailed history / duration of symptoms :

Antenatally Diagnosed or not :

Previous history of respiratory Infections :

(ii) Clinical Examination findings :

(iii) Investigations done : CXR/Fluoroscopy / U/S

C X R : (Finding) :

(iv) Associated anomalies :

(v) Diagnosis :Right/Left/segmental

(vi) Preop Ventilation :

(vii) Management :Operated / Not Operated
:Reason for not operating

(viii) Surgery
1) approach
2) Type of repair
3) Use of ICD
4) Post op ventilation
5) Complication
6) HPE of excised segment

(ix) Long term follow up