

Original Articles

EVENTRATION OF THE DIAPHRAGM: A 10-YEAR EXPERIENCE

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ABSTRACT

Eventration of the diaphragm is a condition characterized by an attenuated but otherwise intact diaphragm.

Twenty patients (age range: 2 days to 11 years) were treated for eventration of the diaphragm during a ten year period (1985-1995). There were 11 girls and 9 boys with a mean gestational age of 37 weeks. 14 patients were born with cephalic presentation, 3 patients with breech delivery and 3 patients by caesarean section. In 5 cases with difficult delivery, forceps were used. Mean birth weight of patients was 2510 gm (range 900 to 3500 g). The most common clinical manifestations of these patients were respiratory distress (16 cases) and failure to thrive (14 cases below 3rd percentile).

50% of patients (10 cases) had from 1 to 5 associated anomalies. Cardiac anomalies were the most common (n=5), followed by respiratory system anomalies (n=4), malrotation (n=3), and hiatal hernia (n=2). All patients had an elevated diaphragm in the chest radiograph (n=20), but only some had paradoxical movement in fluoroscopic studies (n=5). In 8 patients GI contrast studies were performed and three patients were found to have malrotation.

15 patients had right, 3 patients left, and two patients bilateral diaphragmatic eventration. 18 patients underwent operation (plication), while two patients with small and asymptomatic eventration were not operated. Operative repair was performed with no mortality. Hospital stay was 14.5 days. 12/20 patients had come for follow-up and they were evaluated from 5 months to 10 years. All showed good results from diaphragmatic plication without respiratory distress or infection and gained weight from below the 3rd percentile to above the 10th percentile. Surgical correction is strongly recommended in patients with eventration of the diaphragm and failure to thrive or other clinical manifestations.

Keywords: Eventration of the Diaphragm. Bilateral Eventration of the Diaphragm, Plication of the Diaphragm, Respiratory Distress, Failure to Thrive, Cytomegalovirus Infection.

MJIRI, Vol. 11, No. 2, 81-86, 1997.

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INTRODUCTION

Eventration of the diaphragm implies an abnormally high—although intact—diaphragm, arching smoothly from its normal costal attachment.¹ It differs from true diaphragmatic hernia with a sac in that, in eventration, the entire leaf bulges upward from the pressure of the abdominal viscera² whereas in hernia, the diaphragm remains in normal position and the viscera bulge through a localized defect in its surface. Considerable dissatisfaction with the name has been expressed, but no better one has been proposed.²

Eventration can be classified into two groups, congenital (non-paralytic) and acquired (paralytic). Congenital eventration is the result of incomplete development of the muscular portion of the diaphragm.³ Similar to true aplasia, the majority of the diaphragm is replaced by a thin fibrous membrane with a narrow muscular rim attached in the normal position.⁴ The cause of this failure is not known but it has been found in cases of fetal rubella and cytomegalovirus infection³ and in babies with trisomic chromosomal abnormalities.³ In congenital eventration, the phrenic nerve is normal.²

Acquired eventration is usually the result of a traction injury to the roots of the phrenic nerve during traumatic breech delivery, resulting in actual tearing of the roots in severe cases. Paralysis caused by direct pressure on the phrenic nerve by forceps applied to the neck carries a better prognosis.^{3,5} In older infants and children the commonest cause of phrenic nerve palsy is injury during cardiac surgery.⁶

Bilateral congenital eventration of the diaphragm is a relatively rare occurrence, with a high mortality.^{3,4} Congenital eventration occurs more commonly on the left side. Significant herniation *in utero* may produce pulmonary hypoplasia. Other associated anomalies are rare, although trisomy 13, 15 and 18 have been described.^{3,7} Due to positive intraabdominal pressure, the floppy diaphragm is pushed into the chest. The supine position exaggerates the effect. During inspiration, the negative intrathoracic pressure paradoxically increases herniation of the viscera into the ipsilateral chest and the mediastinum to the contralateral side.

There are very few publications concerning eventration of the diaphragm in comparison to congenital diaphragmatic hernia (CDH). There are still controversies about sex incidence, side of eventration, surgical approach and differentiation between CDH and eventration of the diaphragm. Most textbooks and articles do not mention failure to thrive as a clinical manifestation of bilateral eventration, and very little has been said concerning associated anomalies. The need for further research in this field seems obvious.

MATERIALS AND METHODS

The records of patients with a diagnosis of eventration of the diaphragm, treated in the Hospital for Sick Children, Great Ormond Street, London from 1985-1995 were retrospectively reviewed. Data collected included history of delivery (gestational age, normal delivery, breech presentation, use of forceps, caesarean), signs, symptoms and clinical manifestations (cough, wheezing, respiratory distress, apnea, respiratory infection, recurrent respiratory infection, exercise intolerance, vomiting), associated anomalies, imaging studies (chest radiograph, ultrasound and fluoroscopy), side of eventration, kind of incision and operation, suture material, hospital stay and mortality.

Special attention was given to the relationship between weight and clinical manifestations before and after operation on follow up.

RESULTS

There were 20 patients (11 girls and 9 boys) with a female to male ratio of 1.2. Mean gestational age was 37 weeks (range 25 to 42 weeks). The mean age of mothers was 26.8 years (range 21 to 36 years). Forceps were used in four cases with cephalic vaginal delivery and one case with breech delivery (Table I).

Mean birth weight of patients was 2150 g (range 900 to 3800 g), and 4 patients were less than 2 kg. One patient had a history of a previous laparotomy for repair of a left congenital diaphragmatic hernia. This patient had a small right eventration without clinical manifestations and did not need operation.

The most common clinical manifestations of these patients were respiratory distress (16 cases= 80%) and failure to thrive (below the 3rd percentile in 14 cases= 70%) (Table II).

One patient had congenital cytomegalovirus infection. She was born by normal vaginal delivery at 36 weeks gestation, weighing 2100 g. She remained in the neonatal intensive care unit for 10 days. Cytomegalovirus was excreted in urine, nasopharyngeal secretions and sputum. Chest X-ray and ultrasound showed an elevated right hemidiaphragm which moved normally. She had respiratory distress and failure to thrive. Plication of the right hemidiaphragm was performed by a tuck via a posterolateral thoracic approach. She required a chest tube for drainage of right pleural effusion thirteen days postoperatively, and was discharged with medical treatment after recovery.

50% of patients (10 cases) had from one to five associated anomalies (Table III). Cardiac anomalies were the most common (5 patients). Anomalies were more common in male patients (6/9) compared to females (4/11). Overall, 26 anomalies were found in ten patients (6 boys and 4 girls).

Table I. History of delivery in 20 patients with eventration of the diaphragm.

Delivery	No.	%	Forceps No.	%
Cephalic presentation	14	70	4	20
Breech presentation	3	15	1	5
Caesarean	3	15	0	0
Total	20	100	5	25

Table II. Clinical manifestations in 20 patients with eventration of the diaphragm.

Clinical manifestation	No.	%
Respiratory distress	16	80
Failure to thrive	14	70
Wheezing	14	70
Cough	13	65
Respiratory infection	11	55
Recurrent respiratory infection	8	40
Exercise intolerance	3	15
Vomiting	3	15

20 patients (100%) had elevated diaphragms in the chest X-ray, and infection was present in 14 cases (70%). Paradoxical movement was seen on fluoroscopy in 5 patients (25%) and with ultrasound in 3 patients (15%). Upper G.I. contrast study was performed on 8 patients (40%) and 3 of them proved to have malrotation.

18/20 patients (90%) underwent operation (diaphragmatic plication) and 2/20 patients (10%) were managed conservatively. Median age at operation was 104 days (range 2 to 4332 days) and 16 patients (80%) were below 1 year of age (Tables IV, V).

Median weight at operation (18 patients) or at diagnosis (2 patients) was 3.85 kg (range 2 to 43.8 kg). 14 patients (70%) had failure to thrive and were below the 3rd percentile with regard to gestational age (Table V).

15 patients had eventration of the right diaphragm, 3 of the left and two had bilateral eventration (Table VI). Case no. 13, one of twins, had bilateral eventration of the diaphragm, respiratory distress, failure to thrive (below the 3rd percentile), associated anomalies (large ventricular septal defect, stretched patent foramen ovale and congenital heart failure) and nonrotation of the midgut. Nonrotation was corrected at the same time that plication was performed for bilateral eventration. A long transverse upper abdominal incision was given, the diaphragmatic defect was defined, and the anterior rim of muscle sutured to the costal margin bilaterally with interrupted prolene sutures. The cardiac anomaly was operated at another operation. This child is well and is now 5 years old; his weight is between the 10-50 th percentile.

Case no. 14 had bilateral eventration. Operation (bilateral plication) was performed at 3 months of age because of

Table III. Associated anomalies in 20 patients with eventration of the diaphragm.

Cardiac anomalies		
V.S.D.	3	5
Tetralogy of Fallot	1	
A.S.D.	1	
Respiratory system anomalies		
Left congenital diaphragmatic hernia	1	4
Right lung hypoplasia	1	
Pectus excavatum	1	
Upper airway obstruction	1	
Malrotation	3	26
Hiatal hernia	2	
Syndactyly	2	
Dysmorphic child	2	
Myopathy	1	
Klinefelter syndrome (47XXY)	1	
Severe cerebral impairment	1	
Patent vitellointestinal duct	1	
Meckel's diverticulum	1	
Inguinal hernia	1	
Gallstone	1	
Total		

respiratory distress and failure to thrive. During anesthetic induction, problems were encountered during the administration of suxamethonium. The question of myotonia dystrophica was raised. She was electively ventilated for 24 hours postoperatively and then made a good recovery. Muscle ultrasound revealed increased muscle echogenicity consistent with a myopathy, but this was not confirmed by muscle biopsy. Her uncle is susceptible to malignant hyperthermia, and two of his brothers are on the waiting list for muscle biopsy. She is expected to undergo further neurological evaluation.

12 patients (67%) were operated via a thoracic incision, 4 patients (22%) with an abdominal incision and 2 patients (11%) with a long transverse upper abdominal incision. For plication of the diaphragm nonabsorbable monofilament suture material (prolene) was used in all patients. Plication with interrupted sutures was carried out in 11 patients and continuous suture plication in 8 patients (in one case both techniques were used).

Drainage (chest tube insertion) was performed in 3 cases (17%), while 15 patients (83%) were managed without drainage.

Pleural effusion occurred postoperatively in 2 cases (11%) and was treated with chest tube drainage. Recurrence

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of diaphragmatic eventration occurred in one case (5.5%) which did not require reoperation because no clinical manifestations were present.

The mean duration of hospital stay in these patients was 14.5 days (range 3-27 days) and there were no deaths. 12 patients were followed from 5 months to 10 years. Ten patients who had failure to thrive improved postoperatively with weight gain, increasing from below the 3rd percentile to above the 10th percentile within three months of operation. One patient did not have failure to thrive before operation and one patient with bilateral eventration of the diaphragm received no benefit from operation in this regard.

DISCUSSION

The mammalian diaphragm is of complex origin, and not all the details of its development are known. The classic picture of the embryonic components of the diaphragm has been described by Broman who showed that the developing diaphragm receives contribution from the transverse septum, the mediastinum, and the body wall musculature. Closure of the central areas on each side by the pleuroperitoneal membranes completes the diaphragm.² In summary, formation of the diaphragm according to Skandalakis, Gray and Ricketts² is as follows:

1. The pericardioperitoneal canal is a large opening located at the septum transversum on either side of the foregut which is formed because of the inability of the septum transversum to perform a complete separation between thoracic and abdominal cavities.

2. The lung bud is formed within the pericardioperitoneal canal.

3. The thin pleuropericardial fold is a single embryologic entity between the septum transversum and the sternal xiphoid process, which owes its genesis to the ventrally and laterally growing pulmonary expansions.

4. The pleuropericardial fold envelopes the phrenic nerves and the short common cardinal veins before they enter the sinus horn.

5. The pleuropericardial folds fuse together forming the pericardium and pleural cavities.

6. The final step of the genesis of the diaphragm is penetration by myoblasts from the body wall into the membranes of the peripheral part.²

Whatever the precise mechanism, it is the failure of muscularization that leads to eventration. Either the entire diaphragm or only a portion, usually the anterior segment, may be affected. Ravitch and Hendelsman⁸ described such defects surrounded by a ring of muscle in the right diaphragm. In one patient there were two such areas. When the areas are small, the effect is that of a diaphragmatic hernia with a sac rather than of a typical eventration.

The first recognizable description of the condition should

Table IV. Age distribution in 20 patients with eventration of the diaphragm.

<1 month	1 month to 1 year	> 1 year
3 (15%)	13 (65%)	4 (20%)

Table V. Data according to sex, gestational age, age at operation, weight at operation and percentile of weight in 20 patients with eventration of the diaphragm.

Patient Number	Sex	Gestational Age (Weeks)	Age at Operation (Days)	Weight Operation (kg)	Percentile at operation
1	F	31	75	2.54	<3rd
2	F	25	192	4	<3rd
3	M	31	41	2	<3rd
4*	F	40	2	3.14	10th
5	F	41	70	3.30	<3rd
6	F	36	330	5.80	<3rd
7	M	36	664	6.50	<3rd
8	M	39	2	3.15	10th
9	M	40	450	9.30	3rd-10th
10	M	39	28	2.05	<3rd
11	F	40	397	7.80	<3rd
12	F	37	4332	43.80	75th
13	M	38	40	3.10	<3rd
14	F	40	76	3.70	<3rd
15	F	38	263	6.50	<3rd
16*	M	42	262	8.60	10th-50th
17	F	40	178	5.50	<3rd
18	M	38	57	3.20	<3rd
19	F	40	133	6.93	50th-90th
20	M	39	66	3.65	<3rd

* at diagnosis (without operation). F= female, M= male

Table VI. Side of eventration of the diaphragm in 20 patients.

Side	Right	Left	Bilateral
No (%)	15 (75%)	3 (15%)	2 (10%)

be credited to Petit in 1774. Cruveilhier first used the term "eventration".² Morrison⁹ performed the first repair of an eventration of the diaphragm in 1923. Bisgard in 1947¹⁰ performed the first successful operation on an infant.

Eventration can be classified into two groups. Congenital eventration is the result of incomplete development of the muscular portion of the diaphragm. The cause of this failure is not known, but Wayne et al.¹¹ reported an infant girl with bilateral eventration of the diaphragm and congenital cytomegalic inclusion disease. Despite vigorous pulmonary toilet and antibiotic therapy, the bilateral pneumonitis gradually worsened and the patient died on the twelfth day of life. Becroft et al.¹² reported a female infant born with bilateral eventration and prenatal cytomegalovirus infection. She died after 3 hours and 45 minutes. Briggs et al.¹³ reported fetal rubella, and Wexler and Poole¹⁴ trisomic chromosomal anomalies.

Acquired eventration of the diaphragm is usually caused by injury to the phrenic nerve occurring during difficult deliveries with breech presentations. It is produced by a pull on the fifth cervical nerve roots or, in severe cases, there may be an actual tearing of these nerve roots. Less commonly, a paralysis of the phrenic nerve may be due to direct pressure on the nerve caused by delivery forceps applied to the

neck.¹⁵ Stauffer and Rickham in 1972 reported 12 neonates with eventration of the diaphragm secondary to paralysis of the phrenic nerve. All children were born by breech presentation. There were a number of associated lesions, especially Erb's palsy, which occurred in 11 cases. Eight children with paradoxical respiration and marked shifting of the mediastinum were treated by operative plication of the diaphragm and all recovered.¹⁵

In this series only 3 patients were born by breech presentation and one of these with delivery forceps. Four other patients who were born by cephalic presentation had forceps delivery. Paradoxical movement of the diaphragm was seen in one infant with breech presentation and one patient with cephalic presentation. Forceps had been applied in both cases.

A variety of additional malformations have been recorded in cases of eventration, including partial or complete inversion of the stomach and malrotation of the intestines, transposition of abdominal organs, megacolon, hypospadias, pulmonary hypoplasia, defects of the ribs, cardiac anomalies, renal ectopia, cerebral agenesis, hydrocephalus and exomphalos.^{4,7,15,16} Irving and Booker reported the associated anomalies in nine cases and showed that malrotation, cardiac anomalies and renal anomalies were more common.³ From among 20 patients of this series ten (50%) had 1 to 5 anomalies with cardiac, respiratory anomalies and malrotation being more common (Table III).

According to David and Illingworth eventration corresponds to 5% of all diaphragmatic defects.¹⁷ The clinical course of eventration of the diaphragm varies with the extent of involvement. With small unilateral eventration, patients may be asymptomatic or only mildly symptomatic.¹⁸ Many will present with repeated upper respiratory tract infections and have only pulmonary consolidation or a localized diaphragmatic defect visible on the chest radiograph.¹⁹ Respiratory distress, cyanosis, and tachycardia are usual and the stomach may undergo volvulus or may be inverted, with consequent abdominal pain.^{2,3,7}

Malone, Brain, Kiely and Spitz²⁰ reported 22 patients with congenital diaphragmatic defect that presented late, including 4 cases of diaphragmatic eventration. Less than half of the patients had failure to thrive. They emphasize that poor weight gain is a common presenting feature of congenital diaphragmatic defects diagnosed after the neonatal period.

Reynolds²¹ reported that: "in addition to their respiratory distress, these infants suck poorly, become tired during feedings, and fail to gain weight. We have observed infants at 3 months of age who were still at birth weight". In this series 14/20 patients (70%) had failure to thrive (below the 3rd percentile) and we think failure to thrive is a common clinical presentation of diaphragmatic eventration and is an important indication for operation and surgical correction (plication). It should be performed without delay because after operation the child will gain weight. In this series 12/

20 patients were followed up from 5 months to 10 years. Ten patients with failure to thrive improved postoperatively with weight gain from below the third percentile to above the 10th percentile within three months of operation. One patient did not have failure to thrive and one patient who had bilateral eventration had failure to thrive and respiratory symptoms. She recovered from respiratory symptoms but received no benefit with regard to failure to thrive.

Males are more frequently afflicted than females. The ratio has been stated to be as high as 2:1^{22,23} and as low as 4:3. The left side is involved eight times as often as the right.² But in this series 11 of 20 patients are females and the right side is involved five times (15:3) more than the left. We had two cases with bilateral eventration which survived. Reynolds and Amirfaiz advise surgery through the chest for right eventration and through the abdomen for left eventration.^{21,23} We recommend a transthoracic approach for the plication of unilateral (right or left) eventration. A transverse upper abdominal incision is required to facilitate a simultaneous Ladd's procedure or bilateral plication.

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