

EVENTRATION OF DIAPHRAGM IN PEDIATRIC AGE GROUP 10 YEARS EXPERIENCE

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ABSTRACTS

Background: Diaphragm eventration is a congenital condition mostly found in children where there is no diaphragmatic muscle while all the diaphragmatic attachments are normal. Our study was to evaluate the results of early surgical diaphragmatic plication in pediatric age group.

Methods: This descriptive case study was conducted in Thoracic surgery ward Medical Teaching Institute LRH Peshawar Pakistan. All pediatric patients operated for diaphragmatic eventration from June 2008 to May 2018 were analyzed. All those with pediatric age group and having congenital eventration were included in study. Patients with traumatic and malignant eventration were excluded from the study. All the clinical features, demographic data, base line investigations and specific investigations were recorded on a pro forma and analyzed.

Results: Thirty four children were included in our study, who underwent diaphragmatic plication through open thoracotomy in 10 years of study period. Mean age was $8+3.43$ months. male to female ratio was 20(58.8%) were male and 14(41.14%) were female. Twenty eight patients had left sided eventration. Post-operative mean hospital stay was 5.3 ± 2.2 days. The mean follow-up period was 12.3 ± 7.5 months. Clinical symptoms present before surgery were relieved after surgery in 83.3% of children's and unrelieved in 16.7% of children one year after surgery. All children except one new born were alive at the end of one-year follow-up and there was no recurrence of diaphragmatic eventration.

Conclusion: Early diagnosis and open thoracic surgical plication of eventration of diaphragm offers a good clinical outcome with no recurrence.

Key words: Pediatric age group, Diaphragmatic eventration, Plication.

INTRODUCTION

Elevation of diaphragm is usually found accidentally in chest x-ray done in asymptomatic person or some time in patient with respiratory distress. The reason of this elevated diaphragm may be paralysis of diaphragm or congenital diaphragmatic eventration. Eventration is the absence of muscle fibers in between the diaphragmatic membrane congenitally otherwise the anatomical attachments are intact normally.

The muscle tissue is replaced by fibro-elastic tissue histologically which differentiate it from paralysis of diaphragm and hernia of diaphragm.

During autopsies Jean Louis in 1774 first identified the eventration. Beclard in 1829 used the word eventration for the first time. First successful repair of eventration was done by Morrison and was reported in 1923. Incidence of Eventration is a rare and is reported to be $<0.05\%$.

Mostly it is asymptomatic only in 25% patients it is symptomatic with most common symptom is dyspnea. Some patients may present with gastrointestinal

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symptoms like epigastric pain or belching, sometime diaphragmatic rupture or stomach volvulus may be the presenting complications or acute progressive respiratory distress.

Asymptomatic patients do not need treatment. Surgery is the treatment of symptomatic patients, the rationale is that by diaphragmatic plication the vertical diameter of thoracic cavity increases and the abdominal contents will not push the diaphragm upwards during respiration and lying supine.

In children progressive respiratory difficulty and recurrent chest infection should be investigated immediately because early diagnosis of symptomatic eventration and its early surgical management will reduce the post-operative morbidity and mortality with better outcome⁵. Diaphragmatic plication will reduce pressure on the compressed lung, base of the thoracic cavity and mediastinum will also be stabilized and abdominal, thoracic and prothoracic respiratory muscles will be strengthened thus improving respiration⁶.

This study was conducted to evaluate and share our 10 years' experience and symptomatic outcome and recurrence rate of plication for diaphragmatic eventration in infants and children.

MATERIAL AND METHODS

This case a retrospective observational study which was conducted in thoracic Surgery department,

Medical Teaching Institute, LRH, Peshawar, Pakistan. Computerized medical records of patients of pediatric age group who underwent surgery for eventration from June 2008 to May 2018, were reviewed and analyzed. All patients of pediatric age group of either sex, already-diagnosed cases of congenital unilateral diaphragmatic eventration causing symptoms in children were included in the study. Children with paralysis of diaphragm due to other causes like trauma, tumor, neuropathy and myopathy were excluded from the study.

Single lumen endotracheal tubes were used for general anesthesia in all the children who were operated for eventration. Surgery was performed by different surgeons in our unit. Posterior-lateral thoracotomy incision was used and thoracic cavity was opened through 7th or 8th intercostal space. Several U-shaped running sutures were used from posterior aspect of diaphragm to anterior portion of diaphragm with polypropylene suture using Teflon pledgets in between to tighten the diaphragm and increase the vertical diameter of the thoracic cavity. A single intrathoracic drain was put in and wound was closed in layers. After surgery all the children were shifted to the intensive care unit for close monitoring of vitals, children were kept warm and well hydrated.

Clinical presentation, demographic data, site (left or right), CXR findings, cervicothoracic and upper abdominal CT-scan with intravenous contrast, surgical procedure and outcome were noted on a already prepared pro forma. Ultrasound chest and fluoroscopy reports were also analyzed where available. All the follow-up data at one year was noted including clinical features and complications; and compared with preoperative condition.

Thirty days mortality was noted and morbidity was defined as any complication related to the procedure within thirty days like wound infection, pleural space problems, failure of procedure, cardiac, pulmonary and abdominal complications were also documented.

RESULTS

In our study total 34 pediatric patients had thoracotomy performed for diaphragmatic eventration for the past 10 years in our department, out of these 34 children 20 (58.8%) were males and the remaining 14 (41.14%) were females. Mean age of patients was 10 ± 3.43 months. Out of 34 patients 28 (82.35%) patients had left-sided eventration while 6 (17.64%) patients had right-sided eventration. The mean hospital stay was 5.3 ± 2.2 days, and the mean follow-up of our study was 12.3 ± 7.5 months. In 83.3% of children after surgery the pre-operative symptoms were relieved immediately, while in 16.7% of the children symptoms persist till one year follow up after surgery. Patients' demographics and baseline clinical characteristics are given in the table.

In all patients posterior-lateral thoracotomy was performed. Thoracic cavity was opened through 6th or

7th intercostal space. All the patients with left-sided eventration diaphragm was opened laterally with small 3 to 4 cm incision so that the abdominal contents especially stomach could be protected by pulling it down wards so that it could not come in the stich. On right side because of liver diaphragm was not opened.

Plication of diaphragm was done by using polypropylene U-stitches starting from the posterior portion of diaphragm to anterior portion of diaphragm radially and also using Teflon pledgets to ovoid cutting through of diaphragm. Tying these stiches will make the diaphragm firm and tough. A single chest drain was placed in thoracic cavity and the wound was closed in layers.

Post-operatively all the children were kept in intensive care unit. Two of the new born who underwent surgery were kept on ventilator for 24 hours till they recovered fully.

Morbidity in our study was 2(5.88%) including one child developed wound infection and the other developed subcutaneous emphysema. Both of these patients recovered with conservative treatment. One new born child died in our study on first postoperative day (mortality 2.94%) secondary to respiratory failure.

All patients were followed for one year and none of them developed recurrence and all had improved clinically.

Baseline clinical characteristics of eventration in pediatric age group (n=34)

Variable	Frequency
Mean Age	10 + 3.43 months
Male	20 (58.8%)
Female	14 (41.14%)
Left	28 (82.35%)
Right	6 (17.64%)

DISCUSSION

Abnormal and permanent elevation of the hemidiaphragm with normal intact attachments is called eventration of the diaphragm. It is divided into two, one is congenital where there is no muscular development only fibrous tissue membrane and acquired where there is injury of the phrenic nerve⁷. In our study we thoroughly do etiologic investigations of diaphragmatic eventration starting with clinical history, chest radiography PA view, ultrasound chest and CT chest and upper abdomen. In literature the sensitivity of CT chest and upper abdomen is highlighted in the diagnosis of diaphragmatic eventration. We in our study did CT scans in all patients to exclude both neoplastic disease involving the phrenic nerve and sub-diaphragmatic abnormalities (abscess, Chilaiditi syndrome)⁸. In our study, we included only congenital diaphragmatic eventration in pediatric age group patients. Congenital eventration is the result of

incomplete descent of myotomes causing hypoplasia of muscular portion of diaphragm. The exact cause of this failure of descent is not known, but cytomegalovirus infection⁹, fetal rubella¹⁰, and heterozygous genetic mutation with Marfan syndrome¹¹ have been considered to be associated with congenital diaphragmatic eventration.

Different surgical techniques have been discussed in literature to treat congenital diaphragmatic eventration. Lax diaphragmatic excision and re suturing it again, diaphragmatic plication, and placement of prosthesis^{12,13}. Most commonly performed technique is plication of diaphragm through standard thoracotomy. Diaphragmatic plication is done as it has got good results and low complications and low recurrence rate when performed meticulously¹⁴. The aim of plication of diaphragm is to reduce the upwards excursion of the lax diaphragm during inspiration by tightening it with U shaped sutures¹⁵. Our study shows that diaphragmatic plication performed early in infant and children was a safe, simple, and suitable treatment for eventration with good results through a mean follow-up of 12.3 ± 7.5 months.¹²

In present study diaphragm was opened by a 3 to 4cm incision in patients with left sided eventration in order to avoid injury to abdominal organs mainly stomach. Some authors are routinely using this technique,¹² while others avoid opening the diaphragm.⁴ In our view, the diaphragm can usually be opened on left side because sometimes there is pressure of abdominal viscera on diaphragm from below or there are adhesions of abdominal viscera to diaphragm, in such situations opening the diaphragm is safe and should be done.

Comparing our study with other international studies one can find that the symptoms of diaphragmatic eventration have improved dramatically after plication procedure^{5,16,19}. One study by Yazici et al.⁵ shows that diaphragmatic eventration is a condition associated with high mortality so early diagnosis and open surgical plication is the treatment of choice. 33 infants and children were included in his study. Similarly by Obara et al.¹⁶ conducted a study on 18 infants and children, ranging in age from 10 days to 6 years, results of the clinical study suggest that to reduce pathological changes in the lung, early diaphragmatic plication should be done even in patients with acquired diaphragmatic eventration if symptoms of respiratory and digestive system are noted. Tsugawa et al.^[18] concluded in their study that symptomatic patients who have diaphragmatic eventration should have plication done as early as possible to have very good post-operative improvement in their respiratory problems. A study conducted by Jawad et al.^[17] with 8 pediatric patients treated with plication for diaphragmatic eventration included 5 with congenital and 3 with acquired eventration, three of them developed minor postoperative complications and there was no mortality. In one recent study by Wu et al.^[19], where

diaphragm plication surgery in 177 children with a mean age, 10.28 ± 2.35 months was performed in congenital diaphragmatic eventration with no recurrence, and they also concluded that timely diagnosis and early surgical treatment of symptomatic congenital diaphragmatic eventration could effectively resolve respiratory morbidity and reduce complications.

Morbidity in our study was 2 (5.88%) patients. Going through the available literature morbidity associated with plication ranges from 5% to 32% in different series^{12,13,18} but in most studies low morbidity is reported. Some authors have reported some surgery related complications which are rare yet possible like splenic injury¹⁹, abdominal compartment syndrome¹⁸ etc.

Mortality in the study we conducted was one newborn that is (2.94%) who died on second post-operative day secondary respiratory failure the cause of which was severe lobar pneumonia. One mortality also reported by Obara et al.¹⁶ in one patient postoperatively in his series of 18 children that is (5.55%).

CONCLUSION

In our clinical setup early diaphragmatic plication in infants and children is a safe and simple procedure to restore normal pulmonary parenchymal volume by tightening the diaphragm and increasing the anterior posterior diameter of thoracic cavity. There was immediate relief from the symptoms and no recurrence in one year follow up.

REFERENCES

1. Bonanno F. Eventration of the diaphragm: a pitfall in blunt thoracic trauma. *Inj Extra* 2009;40:127-9.
2. Tiryaki T, Livanelioglu Z, Atayurt H. Eventration of the diaphragm. *Asian J Surg* 2006;29:8-10.
3. Shah NN, Mohsin M, Khursheed SQ, Farooq SS, Burch AA, Quraishi AQ. Eventration of diaphragm with gastric volvulus: a case report. *Cases J* 2008;1:404.
4. Groth SS, Andrade RS. Diaphragmatic eventration. *Thorac Surg Clin* 2009;19:511-9.
5. Yazici M, Karaca I, Arik A, et al. Congenital eventration of the diaphragm in children: 25 years' experience in three pediatric surgery centers. *Eur J Pediatr Surg* 2003;13:298-301.
6. Ribet M, Linder JL. Plication of the diaphragm for unilateral eventration or paralysis. *Eur J Cardiothorac Surg* 1992; 6:357-60.
7. Shukla RM, Maitra SK, Patra MP, et al. Eventration of diaphragm with gastric duplication cysts: a rare association. *Indian J Pediatr* 2012;79:1377-9.
8. Mouroux J, Venissac N, Leo F, Alifano M, Guillot F. Surgical treatment of diaphragmatic eventration using video-assisted thoracic surgery: a prospective study. *Ann Thorac Surg* 2005;79:308-12.
9. Becroft DM. Prenatal cytomegalovirus infection and

muscular deficiency (eventration) of the diaphragm. *J Pediatr* 1979; 94:74-5.

10. Briggs VA, Reilly BJ, Loewig K. Lung hypoplasia and membranous diaphragm in the congenital rubella syndrome: a rare case. *J Can Assoc Radiol* 1973;24:126-7.
11. Revencu N, Quenham G, Detaille T, Verellen G, De Paepe A, Verellen-Dumoulin C. Congenital diaphragmatic eventration and bilateral uretero-hydronephrosis in a patient with neonatal Marfan syndrome caused by a mutation in exon 25 of the FBN1 gene and review of the literature. *Eur J Pediatr* 2004;163:33-7.
12. Como JJ, Cohen-Kashi KJ, Alhindawi R. Post-traumatic diaphragmatic eventration. *J Trauma* 2004;56:1149-51.
13. Leo F, Venissac N, Morales F, Rodriguez A, Mouroux J. Plication for diaphragmatic eventration: a simple technique, not a simple problem. *Chest* 2004;125:1170.
14. Celik S, Celik M, Aydemir B, Tunckaya C, Okay T, Dogusoy I. Long-term results of diaphragmatic pli-
cation in adults with unilateral diaphragm paralysis. *J Cardiothorac Surg* 2010;5:111.
15. Groth SS, Andrade RS. Diaphragm plication for eventration or paralysis: a review of the literature. *Ann Thorac Surg* 2010;89:S2146-50.
16. Obara H, Hoshina H, Iwai S, Ito H, Hisano K. Eventration of the diaphragm in infants and children. *Acta Paediatr Scand* 1987;76:654-8.
17. Jawad AJ, al-Sammarai AY, al-Rabeeah A. Eventration of the diaphragm in children. *J R Coll Surg Edinb* 1991;36: 222-4.
18. Tsugawa C, Kimura K, Nishijima E, Muraji T, Yamaguchi M. Diaphragmatic eventration in infants and children: is conservative treatment justified? *J Pediatr Surg* 1997;32: 1643-4.
19. Wu S, Zang N, Zhu J, Pan Z, Wu C. Congenital diaphragmatic eventration in children: 12 years' experience with 177 cases in a single institution. *J Pediatr Surg* 2015; 50.

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