Is Plication for Diaphragmatic Eventration Effective in Improving Lung Function?

44

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Introduction

Diaphragmatic eventration is a rare congenital disorder (incidence <0.05%) that affects the central portion of the diaphragm [1]. It is characterized by a paucity of muscle fibers, while the normal diaphragmatic attachments to the sternum, ribs and spine are not affected [2]. In contrast, diaphragmatic paralysis is an acquired condition in which the muscle fibers might be atrophic, but they are present. Some authors have classified diaphragmatic eventration as congenital or acquired. This is a common misconception, as both pathologies have similar symptoms, physiological impact, and treatment. For this reason, we have chosen to name both diseases "eventration" in this chapter.

Eventration is a diagnosis of exclusion, often asymptomatic in adults, more common in males, and affects predominantly the left hemidiaphragm, although there are cases of right/bilateral involvement reported in the literature. The condition is thought to be caused by abnormal myoblast migration from the third, fourth, and fifth cervical somites into the septum transversum and pleuro-peritoneal membrane [3]. The main symptom that patients experience is dyspnoea, owing to a combination of loss of pulmonary and chest wall compliance and a ventilation/perfusion mismatch [4]. Other non-specific symptoms include epigastric pain, bloating, nausea, and constipation [5]. "Acquired" diaphragm eventration (paralysis) is most

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commonly seen following heart surgery or in pathologies affecting the phrenic nerve such as intrathoracic tumours and neuromuscular disorders. Symptoms are usually worse in children, who sometimes require mechanical ventilation, because they rely more on their diaphragmatic excursion for respiration due to intercostal muscle underdevelopment and a horizontal orientation of the ribs [6].

Surgical repair can be done either open or minimally invasively using a transthoracic or transabdominal approach, and various techniques such as plication, imbrication, and double breasted suturing have been described. Diaphragmatic plication seems to be the most popular method and has the goal to provide symptomatic relief by improving diaphragmatic function; therefore, surgical treatment is reserved exclusively for symptomatic patients [1].

As diaphragmatic dysfunction reduces the compliance of the chest wall, pulmonary function tests (PFTs) can demonstrate a restrictive pattern [4]. Furthermore, the diaphragm plays a critical role in inspiration, therefore measuring the maximum forced inspiratory flow (FIF_{max}) is useful, as is assessing pulmonary function tests (PFTs) in both supine and upright positions (when supine the PFTs decrease between 20% and 50%) [7]. Despite the PFTs often being abnormal in patients with diaphragmatic eventration, these changes are not consistent and do not correlate with the severity of dyspnea.

Surgery for diaphragmatic eventration in children is mostly performed to facilitate weaning off ventilatory support [6], and we have excluded the studies looking at outcomes following plication in this group of patients.

PFTs are useful in monitoring changes after treatment and in providing an objective evaluation of improvement in diaphragm and chest wall function [1]. This chapter reviews the potential benefit of unilateral diaphragm plication in adult patients with unilateral diaphragm eventration in improving lung function tests.

Search Strategy

Electronic searches were performed using the PubMEd, Embase and Cochrane Evidence based medicine databases from 1990 to 2019 and used to identify available data on outcomes after diaphragm plication in adult patients with diaphragmatic eventration published in the English language. The index terms used were "unilateral diaphragm eventration AND plication OR surgery AND outcomes OR quality of life OR physiological changes OR results". Conference abstracts, case reports and studies which included only diaphragmatic paralysis were excluded. The quality of the data was classified using the GRADE system. Table 44.1 shows the PICO terms used to perform the literature search.

Table 44.1 PICO formatted terms for literature search

P(Patients)	I (Intervention)	C (Comparator)	O (Outcome)
Unilateral diaphragm eventration	Plication surgery	Observation	Quality of life, physiologic changes

Results

Most data on outcomes following unilateral diaphragm plication in adults comes from retrospective analyses of case series. Due to the rare nature of this disease, randomized control studies are not feasible, therefore the level of evidence discussed in this chapter is low. Given the myriad of surgical techniques described in the literature, we have divided the results into two sections based on the approach used.

Outcomes After Open Diaphragmatic Plication

In 1992 Ribet et al. [8] published their outcomes after open (thoracotomy) diaphragmatic plication in a cohort of 24 patients over a period of 20 years, including both adults and children, and suggested that one has to be sure the diaphragm is causing the symptoms before performing the procedure (Table 44.2). Out of the 11 adult patients who underwent open diaphragmatic plication, 6 had decreased respiratory function and in 5 cases the post-operative pulmonary function tests were available

Table 44.2 Outcomes after open diaphragmatic plication (thoracotomy)

Author, study type and data collection period	Number of patients	Follow-up period	Pre-operative values	Post-operative values	P value
Ribet [8] retrospective (1968–1988)	11 adults	3 months–18 years	6 patients had decreased respiratory function (no values given)	5 had post-op lung function tests FVC increased by a mean of 20%, FEV1 increased by a mean of 15%	Not given
Calvinho [9] retrospective (1988–2007)	20 adults	4 months–206 months	FEV1% 66.2 ± 15.3 FVC% 70.4 ± 16.0 MRC 2.06 ± 0.97	FEV1% 76.1 ± 20.1 FVC% 78.4 ± 17.3 MRC 1.06 ± 1.14	>0.1 >0.1 0.007
Balci [10] retrospective (2003–2009)	28	12 months	MRC 3.4 ± 0.9 FEV1 1.7 ± 0.6 L	MRC 1.8 ± 0.7 FEV1 2.1 L ± 0.7 L	0.000 0.013
Ali Shah [12] retrospective (2002–2013)	38	6 months	MRC 2.6 ± 0.73 FEV1% 63.5 ± 13.3 FVC% 67.2 ± 14.6	MRC 0.56 ± 0.47 FEV1% 75.2 ± 18.1 FVC% 78.7 ± 12.8	<0.05 <0.05 <0.05
Evman [11] retrospective (2007–2013)	42(23 accordion, 19 double breasted)	12 months	MRC 3 ± 0.6 FEV1% 62 ± 8 FVC% 61 ± 9	MRC 0.9 ± 0.6 FEV1% 76 ± 5 FVC% 76 ± 4	<0.001 No difference between groups

MRC Medical Research Council dyspnea scale, *FEV1* forced expiratory volume in the first second, *FVC* forced vital capacity

and demonstrated an increase in forced vital capacity (FVC) by a mean of 20% and in forced expiratory volume in the first second (FEV1) by a mean of 15%. Interestingly, in 5 cases the diaphragmatic elevation persisted post-operatively, albeit to a lesser extent.

Calvinho et al. [9] retrospectively analyzed outcomes in 20 patients operated between 1988 and 2007 with an average length of follow-up of 59.6 ± 55.1 months. The subjective improvement (dyspnea score) was statistically significant at follow-up (Medical Research Council [MRC] dyspnea scale) improved from a mean of 2.06 ± 0.97 to 1.06 ± 1.14; p = 0.007). Although the PFTs improved post-operatively (FEV1% from 66.2 ± 15.3 to 76.1 ± 20.1 and FVC% from 70.4 ± 16.0 to 78.4 ± 17.3), the differences were not statistically significant. The authors attributed this discrepancy to the small sample size and concluded that subjective improvement is of much more value than the objective data from spirometry.

Balci et al. [10] compared outcomes according to etiology and patch use. In their cohort of 28 patients, 18 cases were secondary to previous operation/disease, 8 were idiopathic, and 2 were post-traumatic. They observed that the patients with a congenital/idiopathic etiology (true eventration) were younger, had a higher diaphragm, were operated sooner after the onset of symptoms, and had better preoperative FEV1 values. In terms of surgical technique, there was no statistically significant difference between open diaphragmatic plication and open diaphragmatic plication and patch. Of note, none of the patients who had the diaphragmatic plication and patch had a post-operative diaphragmatic event; in the plication only group, one patient had an ipsilateral diaphragmatic hernia and one patient developed a recurrence.

Another study by Evman et al. [11] compared mid-term clinical outcomes of open accordion plication vs. open double breasted plication in 42 symptomatic patients with eventration and paralysis. The PFTs and the dyspnea scores (MRC) improved significantly, but there were no statistical differences between the groups. The postoperative increases in FEV1 and FVC were greater than 20% and persisted at 2 years. Although the double breasted technique significantly improved the caudal shift of the diaphragm, this difference did not translate to an increased improvement in pulmonary function tests compared to accordion plication.

Shah et al. [12] retrospectively analyzed 38 patients who underwent surgery for true diaphragmatic eventration over 11 years. They also noted a statistically significant improvement in PFTs and dyspnea score at 6 months post-operatively. Furthermore, they reported a 5.2% 30-day morbidity rate due to surgical emphysema and surgical site infection and a 2.6% 30-day mortality (one patient died due to a fatal arrhythmia).

Outcomes After Minimally Invasive Diaphragmatic Plication

There are only a few studies analyzing outcomes after minimally invasive (VATS or laparoscopy) plication in patients with diaphragmatic eventration. Most are retrospective with the exception of that led by Moroux and colleagues [13], who

Table 44.3 Outcomes after minimally invasive diaphragmatic plication (VATS or laparoscopic)

Author, study type and data collection period	Number of patients	Follow-up period and technique	Pre-operative values	Post-operative values	P value
Moroux et al. [13] prospective (1992–2003)	12	1 year VATs	FVC 1.9 ± 0.8 L FEV1 1.4 ± 0.6 L	FVC 2.47 ± 1.09 L FEV1 1.72 ± 0.8 L	0.0001 0.0006
Groth et al. [14] retrospective (2005–2008)	25	1 year[a] Laparoscopic	SGRQ 59.3 ± 26.8 FVC% 59.2 ± 11.7 FEV1% 55.4 ± 12.9 FIF_{max}% 93.2 ± 34.1	SGRQ 30.8 ± 18.8 FVC% 61.0 ± 10.6 FEV1% 60.9 ± 10.7 FIF_{max}% 115.5 ± 30.9	<0.05 <0.05 <0.05 <0.05
Rombola et al. [15] retrospective (2005–2011)	18	1 year VATS assisted mini-thoracotomy	FVC2.0 ± 0.9 L FEV11.4 ± 0.6 L PEF 5.0 ± 2.0 L PIF 3.4 ± 1.2 L	FVC 2.5 ± 1.1 L FEV1 1.8 ± 0.8 L PEF 5.7 ± 2.0 L PIF 4.0 ± 2.2 L	<0.001 <0.001 0.002 Not significant

FEV1 forced expiratory volume in the first second, *FVC* forced vital capacity, *SGRQ* St. George Respiratory Questionnaire, FIF_{max} maximum forced inspiratory flow, *PEF* peak expiratory flow, *PIF* peak inspiratory flow

[a]Results after excluding data from the patient who resumed smoking

performed VATS plication in 12 patients with unilateral diaphragmatic eventration and prospectively analyzed the results over an 11 year period. Subjective and objective functional improvement as well as radiological improvement were observed in all patients and persisted long term (Table 44.3.).

Groth et al. [14] retrospectively evaluated the short and mid-term results following laparoscopic diaphragmatic plication for symptomatic, unilateral diaphragmatic paralysis and eventration. Their study included 25 patients and showed significant improvement in respiratory quality of life and pulmonary function test results. Unlike other studies, they included the FIFmax in the analysis and found that the improvement 1 year after plication did not reach statistical significance. After investigating the reason behind this discrepancy, they found that one patient resumed smoking after the 1-month postoperative visit. When analyzing the cohort's 1-year PFT data after excluding this patient, they found significant improvements in FVC%, FEV1%, and FIFmax% at 1 year after laparoscopic diaphragmatic plication.

Rombolá and coworkers [15] observed the clinical respiratory and spirometric effects of video-assisted mini-thoracotomy diaphragmatic plication (VAM-TDP) in the treatment of diaphragmatic eventration. Eighteen symptomatic patients were included and significant clinical and spirometric improvement persisted up to 1 year post-operatively. They reported no perioperative mortality, but five of their patients

had postoperative complications (two required non-invasive ventilation, one had a small hepatic hematoma, and two had postoperative ileus).

In summary, both open and minimally invasive approaches yield similar clinical and functional results. The complication rates after open plication are reported as high as 14.3% in one study [10] and 5.2% in another [12], while in minimally invasive plication Groth et al. [14] and Rombola et al. [15] reported that 25% and 27%, respectively, of their cohorts developed a postoperative complication. In contrast, Evman et al. [11] reported no complications in their cohort of 42 patients undergoing open diaphragm plication as did Moroux et al. [13] in their 12 patients who underwent VATS diaphragm plication. The reported complications are similar in all studies and include (but are not limited to) dyspnea, respiratory failure requiring non-invasive ventilation or endotracheal intubation, prolonged drainage, surgical site infections, stroke, and atrial fibrillation. Although the complication rates for minimally invasive diaphragm plication seem to be higher, these results need to be interpreted with care as these studies are retrospective and include different techniques, number of patients, methodology, and reporting.

The post-operative length of stay seems to be similar in both types of approach, ranging between 2 to 15 days across the studies involving open procedures [8–12] and 1 to 15 days in VATs studies [13–15]. However, these results need to be interpreted with caution, as the studies looking into outcomes after open plication involve more patients and have a more variable follow-up period than those studying the outcomes after VATs.

Conclusions and Recommendations

The results of the aforementioned studies should be interpreted with care, as the diagnosis of eventration is often confused with diaphragmatic paralysis. Both open and minimally invasive approaches offer similar clinical and spirometric improvement. However, there is no level 1 evidence comparing the approaches. Diaphragmatic plication is recommended for symptomatic patients with diaphragmatic eventration, as it provides durable improvement in both lung function and symptoms.

Recommendations

- Diaphragmatic plication is indicated in symptomatic patients with diaphragm eventration (evidence quality low; weak recommendation).
- Minimally invasive and open techniques have similar outcomes, thus an approach with which the surgeon is familiar should be adopted (evidence quality low; weak recommendation).

A Personal View of the Data

In our opinion, as with diaphragmatic paralysis, respiratory capacity improvement is likely to occur following repair of eventration at the abdominal-rib cage level in both the operated and contralateral sides, with restoration of synchrony of chest wall movements. These dynamic improvements are not captured by simple spirometry [16]. We prefer a thoracoscopic approach to diaphragmatic plication, as midterm and long term results are similar to other approaches, and the potential for chronic pain is low in our experience. While VATS and laparoscopic approaches have become more popular, a comparative study with open techniques is warranted.

References

1. Groth SS, Andrade RS. Diaphragm plication for eventration or paralysis: a review of the literature. Ann Thorac Surg. 2010;89(6):S2146–50.
2. Christensen P. Eventration of the diaphragm. Thorax. 1959;14:311–9.
3. Schumpelick V, Steinau G, Schluper I, Prescher A. Surgical embryology and anatomy of the diaphragm. Surg Clin North Am. 2000;80(1):213–39.
4. Ridyard JB, Stewart RM. Regional lung function in unilateral diaphragmatic paralysis. Thorax. 1976;31:438–42.
5. Gibson GJ. Diaphragmatic paresis: pathophysiology, clinical features, and investigation. Thorax. 1989;44:960–70.
6. 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(7):1088–92.
7. Clague HW, Hall DR. Effect of posture on lung volume: airway closure and gas exchange in hemidiaphragmatic paralysis. Thorax. 1979;34:523–6.
8. Ribet M, Linder JL. Plication of the diaphragm for unilateral eventration or paralysis. Eur J Cardiothorac Surg. 1992;6:357–60.
9. Calvinho P, Bastos C, Bernardo JE, Eugenio L, Antunes MJ. Diaphragmmatic eventration: long-term follow-up and results of open chest plication. Eur J Cardiothorac Surg. 2009;36:883–7.
10. Balci AE, Ozyurtkan MO. Clinical and surgical specifications of adult unilateral diaphragmatic eventration according to their aetiology in 28 patients. Importance of using diaphragmatic patch and minimal thoracotomy incision. Eur J Cardiothorac Surg. 2010;37:606–12.
11. Evman S, Tezel C, Vayvada M, Kanbur S, et al. Comparison of mid-term clinical outcomes of different surgical approaches in symptomatic diaphragmatic eventration. Ann Thorac Cardiovasc Surg. 2016;22(4):224–9.
12. Shah SZA, Khan SA, Bilal A, Ahmad M, Muhammad G, Khan K, Khan MA. Eventration of diaphragm in adults: eleven years experience. J Ayub Med Coll Abbottabad. 2014;26(4):459–62.
13. 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.
14. Groth SS, Rueth NM, Kast T, D'Cunha J, Kelly RF, Maddaus MA, et al. Laparoscopic diaphragmatic plication for diaphragmatic paralysis and eventration: an objective evaluation of short-term and midterm results. J Thorac Cardiovasc Surg. 2010;139(6):1452–6.
15. Rombolá C, Crespo MG, López PT, Martínez AH, Atance PL, Ramírez AT, Montes JA. Video-assisted minithoracotomy diaphragmatic plication: respiratory effects in adults. Thorac Cardiovasc Surg. 2011;64:647–53.
16. Elshafie G, Acosta J, Aliverti A, Bradley A, Kumar P, Rajesh P, Naidu B. Chest wall mechanics before and after diaphragm plication. J Cardiothorac Surg. 2016;11:25.