Case Report

DIAPHRAGM PPLICATION FOR UNILATERAL DIAPHRAGM PARALYSIS: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Unilateral diaphragm paralysis is an often not recognised cause of dyspnoea.

We present a patient with a unilateral phrenic nerve paralysis treated with diaphragmatic plication. Patient presented with life-style limiting dyspnoea and pulmonary function showed a decrease in FVC when lying down. Since there was no improvement after respiratory muscle training, plication of the hemidiaphragm was performed by a small thoracotomy. The patient improved with regard to respiratory complaints and lung function. Furthermore, we also demonstrate for the first time a significant improvement in exercise capacity with accompanying striking amelioration of quality of life. The patient is now in follow up for more than 2 years and the impressive improvement in exercise capacity remains present.

Surgical treatment of unilateral diaphragm paralysis has been described in case reports and in small series since 1985. Although comparison of the available data is difficult a diaphragm plication seems an effective and safe procedure for patients with symptomatic, acquired unilateral diaphragm paralysis. Improvement of dyspnoea is present in the majority of patients and we even observed an impressive amelioration in exercise capacity. Consequently, it seems appropriate to propose plication to patients with clear symptoms from the moment spontaneous recovery seems unlikely; yet, prospective randomised controlled studies are needed to prove this.

Key words: Diaphragm – paralysis – plication

INTRODUCTION

We present a patient with an acquired unilateral diaphragm paralysis who was treated by diaphragm plication. Postoperative follow up data until 2 years after surgery are presented. To our knowledge, this is the first time that follow up data regarding exercise capacity after diaphragm plication are published. In addition, we present an overview of the available literature.

CASE REPORT

A 61-year-old female patient was referred to our hospital because of progressive dyspnoea. In the past she was diagnosed with asthma, hyperthyroidism and arthritis. She was a former smoker but quit smoking 35 years ago. Her medical treatment consisted of a statin, hormone replacement therapy and glucosamine. She presented with slowly progressive dyspnoea on exertion NYHA class II. Six months earlier, an acute aggravation of the complaints occurred with development of orthopnoea. She couldn't lie down in a complete supine position. Immersion in water aggravated the dyspnoea with a significant intolerance to effort during swimming.

She was obese with a BMI of 30 kg/m². There were no signs of respiratory distress and haemodynamic parameters were normal. Decreased breath sounds were present on the lower part of the right hemithorax. Heart sounds and abdominal examination were normal. Neurological examination showed normal sensibility and normal and symmetrical muscle force of the upper and lower limbs with normal reflexes. Chest X-ray showed an elevated right hemidiaphragm corresponding with the decreased breath sounds.

Spirometry at the time of the first consultation showed a FVC of 99% predicted value and a FEV1 of 81% predicted value, with a decrease of 24% and 26% respectively in supine position. The maximal inspiratory and expiratory muscle forces were – 3.81kPa or 49% predicted value and 8.52 kPa or 69% predicted value respectively, corresponding with a severe limitation of inspiratory and a moderate limitation of expiratory muscle force. Electromyographic examination of the right phrenic nerve showed no diaphragmatic response. Screening of the four limbs was normal. Diaphragmatic fluoroscopy demonstrated a completely immobile right hemidiaphragm with normal respiration and a discrete paradoxical motion during forced in- and expiration, both in upright and supine position. Motion of the left hemidiaphragm was normal.

Based on these results, unilateral diaphragm paralysis was diagnosed and respiratory muscle training was initiated. Nevertheless, symptoms persisted without improvement of pulmonary function or muscle force. Since life-style limiting dyspnoea did not improve, surgery was proposed.

A diaphragm plication by right thoracotomy was performed. A nerve sparing, arch-like incision was made in the middle of the elevated right hemidiaphragm. Afterwards the two parts were sewn back together putting one part over the

other using two rows of separate sutures, thus placing the lateral diaphragm under the medial part.

Postoperatively, the dyspnoea improved significantly with disappearance of orthopnoea and better tolerance to exercise including swimming. Health-related quality of life was evaluated with both the Chronic Respiratory Disease Questionnaire as the SF-36 at baseline and afterwards at 6, 12 and 24 months postoperatively (Table 1). In all 4 categories a higher score was recorded postoperatively with the CRDQ. The SF-36 also showed a clear improvement in quality of life. Not only higher scores were recorded for general health and physical functioning, even mental health, vitality and social functioning improved.

Furthermore, FEV₁ and FVC both in upright and supine position improved significantly and this improvement remained constant over time. Maximal inspiratory and expiratory muscle force also improved. Still not only the measurements at rest improved, also during exercise there was a marked difference. The work rate improved from 120 watt preoperatively to 140 watt at six months postoperatively corresponding with a peak VO₂ of 1581 ml/min or 21.66 ml/min/kg at baseline and 1962 ml/min or 26.88 ml/min/kg at six months. This important improvement in exercise capacity remained constant over the following years (Table 2).

Table 1

	Baseline	6 months	12 months	24 months
CRDQ dyspnea (5-35)	8	28	30	NR
CRDQ fatigue (4-28)	11	14	22	NR
CRDQ emotional (7-49)	33	46	44	NR
CRDQ mastery (7-28)	19	21	26	NR
MRC (0-5)	3	2	2	2
SF36 - physical functioning (0-100)	30	65	60	45
SF36 - roles affected by physical health (0-100)	0	50	100	50
SF36 - roles affected by emotional health (0-100)	100	100	100	100
SF36 - vitality (0-100)	30	30	70	55
SF36 - mental health (0-100)	68	88	92	88
SF36 - social functioning (0-100)	50	100	100	100
SF36 - bodily pain (0-100)	45	45	57.5	45
SF36 - general health (0-100)	45	70	55	60

NR = not recorded.

Table 2

	Baseline	6 months	12 months	24 months
FVC sitting (% pred)	99	122	132	128
FVC lying (% pred)	75	103	113	
TLC (% pred)	92	105	106	104
RV (% pred)	97	100	89	90
MIP (% pred)	55	86	86	90
MEP (% pred)	78	91	80	99
Max work load (Watt) *	123	140	141	140
VO ₂ max (mL/min/kg) *	21.66	26.88	25.27	28.32
VE max *	58.6	60.1	57.6	65.3
Max heart rate/min *	132	133	135	130

NR = not recorded.

DISCUSSION

Unilateral diaphragm paralysis is a clinically important and often not recognised cause of dyspnoea (1). Frequent causes include trauma, neuromuscular disease, infection (herpes zoster, influenza), iatrogenic factors like cardiothoracic surgery, cervical spondylosis and malignancy, although most cases are idiopathic (2, 3).

Patients with unilateral diaphragm paralysis do not have frequent respiratory complaints. Yet, in some there is overt dyspnoea. The main symptom of diaphragm paralysis is dyspnoea on exertion. Changing to supine position and immersion in water tends to increase dyspnoea. Other rather nonspecific, gastro-intestinal symptoms like bloating, heartburn, nausea, regurgitation and epigastric pain also occur (4).

An elevated hemidiaphragm is often an incidental finding on a frontal, upright chest radiograph. This is a very sensitive finding, however not specific enough for the diagnosis of unilateral diaphragmatic paralysis (3, 5). The diagnosis can be confirmed with a "sniff" test during fluoroscopy or ultrasound which shows paradoxical elevation of the paralysed hemidiaphragm during inspiration (2, 3). Computerised tomography of the thorax and upper abdomen is obtained to exclude both a neoplastic disease involving the phrenic nerve and subdiaphragmatic abnormalities (2, 6, 7).

A restrictive lung function pattern is to be expected due to the diminished caudal motion of the affected hemidiaphragm. Vital capacity decreases in the range of 74 to 81% of predicted value (8). Positional decrease in vital capacity is usually more than 10%. In comparison, bilateral diaphragm paralysis is associated with a postural decrease of vital capacity of 25-50% (3, 8).

Electromyography of the phrenic nerve confirms the diagnosis of diaphragm paralysis most definitely and also gives information about the degree of impairment (2, 6).

It also seems sensible to screen for and address the long-term risks of sleep-disordered breathing in symptomatic patients with unilateral diaphragm paralysis because frequent hypopnoeas during rapid eye movement sleep with compensatory activation of the accessory respiratory muscles are observed (9).

Surgical treatment

Diaphragmatic plication can be achieved via the thorax or abdomen, either by open or minimally-invasive surgery. Traditionally a diaphragm plication is done by thoracotomy. Several authors consider a thoracoscopic approach whether or not in combination with a mini-thoracotomy as a safe and effective alternative for conventional surgery with comparable results (6, 10, 11). On the other hand, although incisions are smaller, the incidence and intensity of post-thoracotomy pain does not seem to be very different (12). There might be a higher risk of damaging the very thin paralysed diaphragm and it might be difficult to obtain a sufficiently tense diaphragm using the thoracoscopic technique (1, 2). The short-term results of the minimal invasive approach appear to be promising but long-term data are rather limited and comparative data are lacking (4).

Patient selection is of major importance in order to avoid unnecessary surgery. One has to be certain that the

diaphragm paralysis is causing the symptoms and an underlying neoplastic disease has to be excluded (6). Surgery can only be considered after excluding chronic heart failure and parenchymal lung disease and if symptoms persist, despite physiotherapy and body weight control (2). Spirometry results may not be as important as subjective dyspnoea because there is a poor correlation of dyspnoea score with preoperative pulmonary function tests (7). Most authors advocate a certain observation period before surgery, dependent on the duration of symptoms. This period ranges from 6 to even 24 months (2, 6, 10). However, spontaneous recovery is to be expected in no more than 10% of the cases and seems more frequent after iatrogenic damage of the phrenic nerve during cardiac surgery (13). During the observation period physiotherapy has to be performed and possible weight issues addressed (14). In summary there should be a combination of obvious symptoms and unlikely recovery (1).

Results

Surgical treatment of unilateral diaphragm paralysis had been described in small series since 1985. Comparison of the available data is difficult because it concerns often retrospective data in absence of a control group.

In all series, improvement of dyspnoea in the majority of patients was present and in general patients were satisfied with the postoperative result (Table 2). The majority of the patients who had left their job because of dyspnoea were able to return to work after a couple of months (7, 10). Since different scores to grade dyspnoea were used, comparison is difficult. Freeman describes 4 out of 41 patients who did not realise a significant subjective improvement in their functional status despite significant improvement in pulmonary spirometry. Two of these patients had a body mass index greater than 35. Three of these patients also had been known to have a paralysed hemidiaphragm longer than 4 years before plication. Intuitively it seems that these factors may influence the outcome of patients undergoing diaphragm plication but the power of the study was not sufficient to draw firm conclusions (10).

In our case there was not only a grading of the dyspnoea by the MRC/ATS grading system or the visual analogue scale as was the case in most series, but also the health-related quality of life was evaluated with both the Chronic Respiratory Disease Questionnaire as the SF-36 with proof of amelioration on all questionnaires. CRDQ examines four aspects of patients' lives: dyspnoea, fatigue, emotion, and mastery (the feeling of control over the disease and its effects). SF-36 includes eight health concepts, each having a 0-100 scoring range: physical functioning, roles affected by physical health, bodily pain, general health, vitality, social functioning, roles affected by emotional health, and mental health. Although it is more time-consuming to incorporate these kind of questionnaires, it might be possible to detect differences in post-operative status as compared with before that are more important in daily life, more so than when only focusing on the dyspnoea as such.

In the different series all spirometric values seemed to improve postoperatively (Table 3). In our case the FVC and FEV1 (and TLC), although still within normal limits at baseline, also increased significantly. Versteegh et al observed the typical deterioration when changing from upright to supine

Table 3

Author (ref)	Year of publication	N	Follow up	% improvement FEV ₁	% improvement FVC	Complications
Wright (20)	1985	7	NR	NR	NR	NR
Graham (21)	1990	17	≥ 6 weeks	NR	18,5	NR
		6	5 years (range 4-7)		17,2	
Ribet (19)	1992	11	8,5 years (range 3 months-18 years)	14	16	NR
Higgs (12)	2002	19	10 years (range 7-14)	15,4	11,8	NR
Mouroux (11)	2005	10	1 year	22,9	30	pneumonia (n=1)
		6	5 years	25	18,3	
Freeman (15)	2006	25	6 months	21,4	17	DVT (n=1)
						superficial wound infection (n=1)
Versteegh (1)	2007	22	4,9 years (range 1,2 - 8,7)	7	9	death (n=3)
						paralytic ileus (n=5)
Freeman (16)	2009	41	6 months	23	19	pneumonia (n=2)
			4 years	21	17	atrial fibrillation (n=2)
						prolonged ileus (n=1)
						DVT (n=1)
Celik (3)	2010	13	5,4 years (range 4-7)	27,3 ± 10,9	43 ± 30,6	death (n=1)
						superficial wound infection (n=1)

NR = not recorded.

DVT = deep venous thrombosis.

position. After surgery there was an obvious and significant decrease of this effect both for the loss of VC as for loss of FEV₁ (1). Graham had comparable results (14). In contrast with these results Higgs et al. found only a limited effect of positional changes on pulmonary function, both before and after surgery. And surgery did not significantly alter postural change in lung volume (7). It seems logical that there is an improvement in FVC by repositioning the diaphragm, still the muscle remains paralysed and an important question is whether the surgery corresponds with an objective functional improvement.

To our knowledge, this is the first time that follow-up data regarding exercise capacity after diaphragm plication are published. The improvement in peak VO₂ is not only significant but stays constant during follow-up. Measuring exercise capacity during ergospirometry probably translates better the way a patient functions and an improvement in peak VO₂ might be more significant in daily life than a normal position of the diaphragm on chest X-ray or an improvement in FVC. We believe that if this improvement in peak VO₂ can also be measured in a series of patients, this further supports the use of surgery for the treatment of diaphragm paralysis.

Different studies presented long-term data concerning a small group of patients. All show that the positive effect of surgery occurred early postoperative and was sustained over time (6, 7). This was also true in our patient. Freeman was the first to use a minimal follow up of 48 month as one of the inclusion criteria. In this series FVC, FEV₁, FRC and TLC improved significantly and this improvement remained constant over time (10). Celik et al also had a mean follow-up of 5.4 years (range 4-7). Mean FVC and FEV₁ improved significantly at late follow-up and also MRC/ATS dyspnoea scores improved significantly (2). In rare cases a slight elevation of the plicated diaphragm becomes visible on X-ray. Nevertheless

most of the time this elevation is very limited and asymptomatic. Only one patient required a re-plication (1, 2, 7, 11).

Diaphragm plication seems to be a safe procedure with limited morbidity and mortality. Possible perioperative complications are those of thoracotomy in general such as pneumonia, atrial fibrillation, prolonged ileus and deep venous thrombosis. Only 2 of the authors mention immediate in-hospital deaths. Celik described one patient who died of postoperative sepsis, in the series of Versteegh there were 3 deaths, 1 who died of an acute myocardial infarction, 1 because of pulmonary embolism and the third one because of the combination of renal and right cardiac failure (1, 2). In none of the series there is long-term mortality attributable to the surgery (1, 2, 6, 7, 10).

A serious limitation of all the current reports is the lack of a control group. In a very limited study about bilateral diaphragm paralysis 3 patients were operated on and 3 were observed. During follow up all patients in the control group deteriorated (16). Freeman described in his first series that despite continued pulmonary rehabilitation, patients who declined diaphragm plication experienced no significant change in their mean pulmonary spirometry or MRC scores at their 6-month follow-up visit. There was a trend, however, toward worsening of these values (15).

CONCLUSION

Diaphragm plication is an effective and safe procedure for patients with symptomatic, acquired unilateral diaphragm paralysis. Follow-up shows not only an improvement in pulmonary function but also a clear decrease of subjective dyspnoea. Moreover, in our case, we showed a benefit on exercise capacity and overt amelioration in quality of life,

which remained present during the follow-up period of 2 years.

Consequently and especially since there exists no alternative treatment, it seems appropriate to propose plication to patients with clear symptoms from the moment spontaneous recovery seems unlikely. However, randomised controlled trials to clearly demonstrate the effects of plication are needed.

CONFLICT OF INTEREST: None.

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