Study objectives: To assess the feasibility and clinical outcome of bilateral plication of the diaphragm in patients with bilateral diaphragmatic paralysis (BDP) caused by neuralgic amyotrophy (NA), a mononeuritis of the phrenic nerves.

Design: Prospective, case-control study over a 1-year period.

Setting: A university hospital in The Netherlands.

Patients: Six patients who presented with BDP caused by NA.

Methods: The diagnosis of BDP was based on the absence of muscle response after cervical magnetic stimulation of both phrenic nerves. Three patients did not undergo surgery but were observed for a period of 2 years, and the other three patients underwent a limited lateral thoracotomy at the eighth intercostal space. Plication was performed by U-stitches until the diaphragm was as tight as possible. Vital capacity (VC) and arterial blood gas was measured during follow-up.

Results: One month postoperatively, mean VC measured in the supine position was significantly improved by 17%, and this effect was sustained for 12 months. Arterial Po2 increased by 45%. VC and blood gas levels did not improve in the three patients that were only observed during the 2-year period. All three surgical patients could sleep in the supine position after the operation.

Conclusion: Bilateral plication of the diaphragm for NA-induced paralysis results in improvement of ventilation and blood gas exchange, allowing patients to sleep in the supine position without dyspnea.

Key words: diaphragm; neurogenic amyotrophy paralysis; plication

Abbreviations: BDP = bilateral diaphragmatic paralysis; MIPm = maximal inspiratory mouth pressure; NA = neuralgic amyotrophy; VC = vital capacity

Patients with bilateral diaphragm paralysis present with severe breathlessness and the inability to sleep in the supine position. The condition may deteriorate into cor pulmonale. Paralysis of the phrenic nerves may be caused by nerve compression, vasculitides, or neuromuscular diseases, including Pompe’s disease.1 When none of these conditions are present and when bilateral diaphragmatic paralysis (BDP) coincides with acute severe pain localized in the shoulder region, the phrenic paralysis may be diagnosed as neuralgic amyotrophy (NA).2,3 We recently identified six patients with BDP due to this condition. They were referred to our clinic for treatment of their severe respiratory failure. In the literature, conventional treatment of NA consists exclusively of administering analgesics.2,3 A poor prognosis is reported in several case studies.4,5 Plication of the diaphragm to decrease lung compression induced by unilateral phrenic paralysis is reported as an effective treatment for dyspnea.6 To our knowledge, the results of plication in patients with BDP have not been published. Our patients with BDP due to NA had serious respiratory failure and, therefore, a poor prognosis for survival. We now report the clinical effect of bilateral plication of the diaphragm in three of these patients.

Materials and Methods

Study Subjects

Six patients who had isolated BDP caused by NA were assessed. The patient characteristics are presented in Table 1. All six patients had signs of right-sided heart failure. They were unable to lie flat and could sleep only while sitting in a chair. A neurologist found no signs of limb, neck, or thoracic muscle paralysis. Cervical magnetic stimulation and bilateral electrical stimulation of the phrenic nerves showed no activity in any of the
patients. CT scans of the cervical and thoracic part of the phrenic nerve could not be performed because the patients could not lie flat.

**Pulmonary Function Tests**

Preoperative vital capacity (VC) was recorded using a dry rolling-seal spirometer (Morgan Spiroflow; Rainham, UK) according to standard recommendations. Patients were positioned individually as flat as possible allowing the VC maneuver to be performed. As a result, the VCs were measured with each patient lying in a different position. VC also was recorded while patients were in the upright position. At follow-up, VC was recorded in the supine and upright position.

Maximal inspiratory mouth pressure (MIPm) was recorded using a mouthpiece and tube assembly while patients were sitting in a chair. MIPm was measured by asking the subject to exhale to functional residual capacity, at which point a valve in the assembly tube was closed and the subject inhaled as vigorously as possible. Measurements were recorded three times, and the best value was expressed as a percentage of the predicted value according to Black and Hyatt.

Arterial blood gases were drawn from the radial artery and were processed immediately using a blood gas analyzer (model 1312 Blood Gas Manager; Instrumentation Laboratory Inc; Lexington, MA). Right-sided heart catheterization was performed at rest as described by Grossman and Baim.

**Surgical Procedure**

Three patients underwent surgery. The procedure was performed bilaterally through a limited lateral thoracotomy in the eighth intercostal space. Both sides of the diaphragm were operated on during the same procedure. In all cases, the diaphragm was very thin, with the contours of organs such as spleen, stomach, and colon clearly visible. The uncut diaphragm was plicated with a number of parallel U-stitches (Mersilene 2; Ethicon; Norderstedt, Germany), starting in the middle close to the attachment of the pulmonary ligament, until the diaphragm stiffened as much as possible (Fig 1). From there, plication was continued toward the mid-axillary line, resulting in a large part of the plicated tissue being located in the central tendon. However, in patients in whom the paralysis had existed for a long time, such as in case 4, the amount of redundant tissue was large, requiring muscle plication as well.

**Statistical Analysis**

Significant differences between the preoperative and postoperative measurements of VC, MIPm, and arterial blood gases were calculated with a Student’s t test.

**RESULTS**

The results of pulmonary function tests administered to patients at presentation in our clinic are shown in Table 1. The pulmonary artery pressures (mean ± SEM) were elevated in all six patients (systolic, 62 ± 15 mm Hg; diastolic, 46 ± 8 mm Hg). Patients 1 and 2 were the first patients in whom disease was diagnosed, and at that time we recommended only oxygen supplementation for relief of symptoms. Patient 3 (Table 1) refused the proposed operation. The three patients on whom surgery was performed were extubated immediately after the operation. All three patients could lie in the supine position within 2 days after the operation. For a period of 3 months after the operation, all three patients experienced tightness in the region of the xyphoid process. Within 6 months after the operation, the two male patients could resume their full-time jobs.

Table 2 shows the results of follow-up of all six patients. In patients who underwent surgery, the

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**Table 1—Patient Characteristics*  

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age, yr</th>
<th>Height, cm</th>
<th>Weight, kg</th>
<th>VC, % predicted</th>
<th>MIPm, % predicted</th>
<th>PaO₂, mm Hg</th>
<th>PaCO₂, mm Hg</th>
<th>pH</th>
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</thead>
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<tr>
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<td>60</td>
<td>45</td>
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<td>63</td>
<td>50</td>
<td>63</td>
<td>43</td>
<td>7.41</td>
</tr>
</tbody>
</table>

*Patient did not undergo surgery on their diaphragm.

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**Figure 1.** A number of U-stitches were used to plicate the diaphragm as tightly as possible in both the lateral and anteroposterior direction. Thereafter, the redundant tissue from the center was used to cover the folds and to add additional tension to the diaphragm. This procedure was performed with a running suture (Ethibond 2/0; Ethicon).
mean VC measured while in the supine position significantly improved from 51% of the predicted value to 68% (p < 0.01). Mean arterial oxygenation improved from 59.2 to 93.4 mm Hg (p < 0.01), while MIPm did not significantly change. Patients 1 to 3, who did not undergo surgery, had no clinically meaningful changes in lung function parameters.

**DISCUSSION**

Diaphragmatic paralysis producing symptoms in adults is an uncommon clinical problem, with sparse literature available about its treatment and long-term outcome. After observing the first two patients, who remained in poor clinical condition, we were particularly struck by the third patient (patient 4 in Table 1). This patient was a 47-year-old police officer who worked in active service on the street and who presented with signs of severe pulmonary hypertension that had developed in 2 weeks time, together with aching pain in neck and shoulder. Although oxygen supplementation relieved his dyspnea, his clinical condition deteriorated significantly over a period of 3 months. In the meantime, we found out that 23 years prior to his present admission he had experienced left-sided phrenic paralysis (Fig 2, top). Thus, it appeared that this patient presented with acute, unilateral, right-sided phrenic paralysis with a preexisting paralysis on the left side. Based on positive results from the plication of unilateral diaphragm paralysis, it was decided to offer him bilateral plication. This procedure resulted in an excellent clinical result. Thereafter, we offered the operation to four other patients of whom one refused (patient 3 in Table 1).

It has been reported that relief of symptoms and improvement of pulmonary function is sustained for ≥5 years after unilateral plication.⁶ No data are available for BDP, but the results reported in Table 2 are comparable to the initial results obtained 1 year after plication for unilateral diaphragm paralysis that were reported by Wright et al.¹⁰ NA of the phrenic nerves was diagnosed our patients. This diagnosis was obtained after the exclusion of other diseases such as nerve compression, vasculitides, and neuromuscular disorders. In theory, the absence of diaphragmatic activity during cervical magnetic stimulation may not be regarded as final proof for the existence of NA, since it cannot be ruled out that the phrenic nerves were not hit during the procedure. Nevertheless, all our patients had symptoms similar to those described by Mulvey et al.³ with pulmonary function tests showing reduced FVC in the supine position compared to the upright position and hypoxemia with normal diffusing capacity of the lung for carbon monoxide (carbon monoxide data not shown). Consequently, a diagnosis of NA should be considered in patients with diaphragmatic paralysis even in the absence of upper limb weakness. It is possible that the incidence of phrenic involvement in NA may have been underestimated in the past and that NA may account for more cases of hemidiaphragmatic paralysis followed by a contralateral episode leading to BDP than previously has been recognized.³ It has been reported that likelihood of recovery is smaller if bilateral phrenic involvement occurs on the initial presentation of a patient with NA.³ Of 12 adult patients reported by Mulvey et al,³ all but 1 patient improved symptomatically, but there was no consistent change in lung volumes, diaphragmatic function, or global inspiratory strength during a follow-up period of 2 to 4 years. Our patients who did not undergo bilateral plication showed a similar pattern.

A study of experimental surgery in dogs with BDP, induced by phrenicotomy with an open chest, found that bilateral plication of the diaphragm did not improve lung compliance and the work of breath, except for tidal volume.¹¹ The results of this study in dogs supported the observations found after bilateral

| Patient | VC in Supine Position | | PaO₂ | | MIPm |
|---------|-----------------------|---|-----|---|-----|---|
|         | % Predicted | Change, % | mm Hg | Change, mm Hg | % Predicted | Change, % |
| Follow-up without surgery | | | | | | |
| 1       | 42              | 5   | 64.6 | 7.6 | 30 | 6   |
| 2       | 55              | 5   | 67.6 | 3.8 | 43 | −2  |
| 3       | 55              | −3  | 60.8 | 6.0 | 44 | 4   |
| Follow-up with surgery | | | | | | |
| 4       | 56              | 16  | 91.1 | 38.7 | 35 | 5   |
| 5       | 88              | 20  | 101  | 38.7 | 43 | 3   |
| 6       | 61              | 14  | 88   | 25  | 47 | −3  |

*Difference in values at 12 months minus values at presentation.*
The application of the diaphragm in infants, in whom this treatment was not as effective as unilateral plication, possibly due to weak rib cage muscles. The results of our case-control study are in contrast to these results, possibly due to differences in chest wall mechanics. In this respect, it is of importance to notice that the results of our MIPm measurements indicated that diaphragm plication caused no significant change of inspiratory muscle effort after surgery. We were concerned that plication might compromise an adaption mechanism to BDP in the upper rib cage and in the sternomastoid and trapezius muscles, however, our MIPm data did not support the presence of such an effect.

In conclusion, our data show that diaphragmatic plication for BDP in adults improves pulmonary function and gas exchange and makes it possible for patients to return to their normal daily activities.

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REFERENCES


Figure 2. Top: chest radiograph of patient 4 made at application for military service 23 years before he presented to our clinic. The left diaphragm is elevated to the level of the left hilum. Middle: chest radiograph shows the bilateral elevation of the diaphragm at presentation in our clinic with both diaphragms almost reaching the hila. Bottom: chest radiograph shows increased lung volumes at inspiration 12 months after bilateral plication.