hemolysis was identified in patients who received lyophilized IVIG products with low anti-A/B titers.

The administration of IVIG for the preparation of patients with MG before thymectomy should follow a careful risk-benefit analysis in each patient and lead to monitoring of hemoglobin levels after IVIG administration and before operation; this did not occur in our patient. The transfusion of blood should be avoided during active hemolysis; however, critical postoperative anemia should be treated with leukocyte-reduced type O blood, avoiding type A or B blood. The surgeon responsible for the care of patients with MG should participate in decisions regarding their preoperative management because he or she will manage any postoperative complications. Close monitoring of hemoglobin 48 to 72 hours after IVIG infusion and before operation is advised, with repeated testing for symptoms of anemia at least 2 weeks thereafter. If anemia is identified, a hemolytic workup is recommended, including a direct antiglobulin test, smear analysis, determination of lactate dehydrogenase and haptoglobin levels, and a reticulocyte index. Hemolysis may be avoided by choosing a low-titer product and should be considered in patients with a non-O blood group treated with high-dose IVIG who would not tolerate anemia.

References


Dramatic Improvement After Bilateral Diaphragmatic Plication in Charcot-Marie-Tooth disease

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A 52-year-old woman with Charcot-Marie-Tooth disease presented with severe dyspnea due to bilateral diaphragmatic paralysis severely compromising respiratory function. There was little in the available literature to guide us regarding management of this unusual condition, and after deliberation, we decided to treat her with a staged plication of bilateral hemidiaphragms. Postoperatively, she demonstrated very good symptomatic relief supported by objective evidence, including improvement in lung function tests. We describe our management of this difficult condition, including the surgical and anesthetic considerations, and would recommend bilateral diaphragmatic plication as an effective option in patients with this unfortunate disease.

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A pulmonary function test (PFT) revealed severely compromised respiratory function, with a forced expiratory volume in 1 second (FEV1) of 30% of predicted and a forced vital capacity (FVC) of 33% of predicted. The patient was keen for some relief from her symptoms. The option of nocturnal continuous positive airway pressure therapy or staged diaphragmatic plication was discussed with the patient, and a surgical approach was agreed upon. We were unable to find much literature to guide us on how to manage this difficult condition.

Before the initial plication, arterial blood gas analysis revealed type 2 respiratory failure, with pH of 7.41, partial pressure of oxygen of 7.9 kPa, partial pressure of carbon dioxide of 7.1 kPa, and HCO₃ of 33.9 mmol/L.

A general anesthetic was administered. The use of regional anesthesia has been described for obstetric and orthopedic procedures, without exacerbating the underlying disease process, in this cohort of patients [1]. However, we avoided an epidural because of the risk of impairing intercostal muscle function postoperatively as our patient was reliant on the integrity of these muscles for ventilation. Intrathecal morphine was avoided because of the risk of causing unpredictable respiratory depression. Epipleural and wound catheters infused plain 0.25% bupivacaine by specialized pumps, in conjunction with simple analgesics.

A limited right posterolateral thoracotomy through the seventh intercostal space was made. The diaphragmatic muscle was atrophic. The central tendon of the right diaphragm was identified, and 4 pairs of pledgedet No. 5 Ethibond (Ethicon, Somerville, NJ) sutures were used to plicate the diaphragm using a horizontal mattress technique. Intermittent noninvasive respiratory support was required for the first postoperative night. The patient’s postoperative recovery was uncomplicated, and she was discharged home on postoperative day 6.

She was reviewed after 6 weeks, at which time she reported very good improvement in symptoms, and her use of home oxygen had become less frequent. A repeat PFT showed improvement in the FEV₁ to 41% and FVC to 46%, with a FEV₁/FVC ratio of 76% (Table 1). Blood gases had improved, revealing pH of 7.42, partial pressure of oxygen of 9.48 kPa; partial pressure of carbon dioxide of 6.28 kPa; and HCO₃ of 30.2 mmol/L. Encouraged by the results, we decided to proceed with plication of the contralateral side.

Intrathecal preservative-free morphine was used because there was marked symptomatic improvement along with objective evidence from the PFT. On this occasion, we avoided muscle relaxants after induction of general anesthesia because residual effects might have contributed to the patient’s deterioration in respiratory function postoperatively.

A similar surgical procedure was performed on the left side through an eighth intercostal space minithoracotomy. The patient was discharged home on postoperative day 5. She was followed up at 5 weeks and reported dramatic improvement. She was now able to lie flat and did not require any supplemental oxygen. A chest roentgenogram showed near-normal appearance of the diaphragms. A PFT at this stage demonstrated the FEV₁ was 51% and FVC was 53%, with an FEV₁/FVC of 82% (Table 1).

**Comment**

Charcot-Marie-Tooth disease is the commonest inherited neuromuscular disorder, with an incidence of 1 in 2,500 [2]. It is a group of disorders causing muscle wasting, weakness, and sensory loss and can cause restrictive pulmonary impairment in association with phrenic nerve dysfunction, diaphragm dysfunction, or thoracic cage abnormalities [3]. The efficacy of plication for diaphragmatic paralysis has been previously proven [4, 5]. The difficulty in Charcot-Marie-Tooth is that some of the

<table>
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<tr>
<th>Variable</th>
<th>Preoperative</th>
<th>Right</th>
<th>Left</th>
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<tbody>
<tr>
<td>FEV₁ %</td>
<td>30</td>
<td>41</td>
<td>51</td>
</tr>
<tr>
<td>FVC %</td>
<td>33</td>
<td>46</td>
<td>53</td>
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FEV₁ % = forced expiratory volume in 1 second, percentage of predicted; FVC % = forced vital capacity, percentage of predicted.
respiratory compromise is related to the wasted thoracic muscles.

When faced with this patient, we were unsure about the extent of contribution to the symptoms related to the diaphragm paralysis. Initially, we were keen to try noninvasive ventilation as a treatment strategy. After some deliberation, we decided to try plication as mainly a method by which the patient could be more able to recline. We were pleasantly surprised by the improvements in the patient’s symptoms and by the objective improvement seen in the PFT results.

The staged approach allowed not only for stabilization of the patient but was also helpful in assessing improvement. We present this case to highlight the point that bilateral plication of paralysed diaphragm in Charcot-Marie-Tooth disease can lead to a dramatic improvement in quality of life, supported by objective evidence, and should be considered in this subgroup of patients.

References

Thoracoscopy-Assisted Minimally Invasive Surgical Stabilization of the Anterolateral Flail Chest Using Nuss Bars

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Flail chest is caused by complex fractures of multiple ribs as a result of severe chest injuries, which results in paradoxical chest movements that severely compromise respiratory function. We report our experience of thoracoscopically assisted, minimally invasive surgical stabilization of massive anterolateral flail chest using a Nuss bar in three patients. This technique offers effective stabilization while having the advantages of short surgical time, minimal blood loss, less trauma, quicker recovery, and small and inconspicuous incisions.

Flail chest is caused by complex fractures of multiple ribs as a result of severe chest injuries. Because of the paradoxical movement, a flail chest can cause a significant decrease in tidal volume and an inability to clear airway secretions. These patients are prone to developing pneumonia and respiratory failure. Traditionally, patients with flail chest are treated with prolonged mechanical ventilation. In recent years, surgical fixation of the flail segment has become more accepted in selected patients. Several studies have shown that surgically treated patients have decreased ventilation and ICU requirements, a lower risk for pneumonia and mortality, less residual chest wall deformity, reduced total cost of care, and quicker return to work [1, 2]. Despite these advantages, surgical stabilization of flail chest remains underutilized because of the unfamiliarity of surgeons with stabilization techniques and the lack of effective hardware [3, 4].

We report our experience with thoracoscopically assisted, minimally invasive surgical stabilization of massive anterolateral flail chest using a Nuss bar in three patients. This technique offers effective stabilization while having the advantages of less trauma and quicker recovery.

Case Reports
Surgical Technique

Chest measurements were performed before surgery, and steel bars of appropriate length were selected. Steel bars were bent using anvil bar benders according to the threedimensional reconstruction of their CT images. Using general anesthesia, a 1.0-cm incision was made in each uninjured chest wall on both sides of the flail segment. The pleural cavities were inspected using video-assisted thoracoscopy. Blood clots were removed and the pleural cavities were irrigated. Submuscular tunnels were made from each incision to a position lateral to the flail segment. A Nuss bar (Pectus Support Bar System; PTY Medical Device, Shanghai, China) was passed from a pre-selected uninjured intercostal incision, through the submuscular tunnel, and into the pleural cavity under thoroscopic monitoring. The Nuss bar was passed across the medias- tinum under the flail segment, and we brought out the chest wall through a preselected, uninjured intercostal incision through the submuscular tunnel, and into the pleural cavity under thoroscopic monitoring. The Nuss bar was rotated 180 degrees so that the convexity of the bar faced anterior to support the flail segment. The two ends of the bar rode on the uninjured ribs with the mid portion of the bar placed behind the flail chest wall. A second bar was placed as needed to support larger flail segments. The patients were observed in the intensive care unit with adequate analgesics and antibiotics. Once fracture healing was demonstrated on follow-up chest radiographs, the Nuss bars were removed in the operating room using general anesthesia.

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