Pneumothorax in chronically ventilated neuromuscular and chest wall restricted patients: A case series

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INTRODUCTION: Pneumothorax occurs in chronically ventilated neuromuscular and chest wall-restricted patients. Non-invasive home mechanical ventilation (HMV) and airway clearance techniques are standard care, but are perceived to put patients at increased risk of pneumothorax. Malnutrition as reflected by low body mass index was identified as an association with pneumothorax in this case series.

METHODS: Retrospective chart review of six cases of pneumothorax in patients with neuromuscular and chest wall diseases receiving chronic HMV.

RESULTS: Six cases were reviewed. Median duration of HMV at time of the pneumothorax was 3 years (range 0.25 to 9 years). Five patients used mechanical ventilation guidelines (4). One of six patients reported that she was not actually using LVR prior to presentation with pneumothorax; however the remaining five patients did use airway clearance strategies on a regular basis (Table 1).

Pneumothorax characteristics

Three patients had small pneumothorax (<2 cm) on initial presentation and three had large pneumothorax (>2 cm) using the BTS guidelines definition for spontaneous pneumothorax (5).

CASE REPORTS

Case 1

A 61-year-old male with ALS presented with a right pneumothorax (Figure 1A). The patient was treated with a chest drain and upon removal of the tube, a small loculated basilar right pneumothorax persisted, as documented on CT (Figure 1B). The patient was ventilator-dependent at night, and continued using NIV throughout his treatment for pneumothorax. Airway clearance maneuvers (LVR) were held until radiographic stability was achieved on CT of the chest.

Figure 1A) Chest xray of ALS patient at presentation with large right pneumothorax

CASE REPORTS

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Case 2
A 24-year-old female with limb girdle dystrophy presented with a sentinel event, a tiny left apical pneumothorax (Figure 2A) while using nocturnal NIV (BPAP 18/10) and LVR. This was accompanied by symptoms of increased dyspnea and orthopnea that had started abruptly in the days prior to assessment. No chest tube was placed at that time, but LVR was suspended. Two months later, the patient represented with a large right pneumothorax (Figure 2B). A chest tube was required. After hospital discharge one month later the patient had a large left pneumothorax. Surgical pleurodesis was considered but declined by the patient. Another pneumothorax on the right occurred two months later and pigtail chest tube was inserted. A doxycycline pleurodesis was performed on the right at that time. This procedure was well-tolerated although precautions were taken by admitting the patient to intensive care and intubating her prior to pleurodesis (Figure 2C). The patient was successfully extubated and no subsequent pneumothorax occurred.

Case 3
A 53-year-old female with CAP myopathy and coexisting anorexia and anxiety disorder developed progressive respiratory failure with high level NIV dependence (22 hours per day). The patient had been prescribed but was not using lung volume recruitment. After a change in respiratory status, which was subsequently felt to be related to an aspiration event, the patient underwent a CT scan (Figure 3). The patient was found to have a <1 cm apical right pneumothorax and pneumomediastinum. Management was conservative; NIV was continued, with no chest tube drainage. There was no recurrence of pneumothorax in this case.

**TABLE 1**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Gender</th>
<th>Diagnosis</th>
<th>FVC (L)</th>
<th>Wt (kg)</th>
<th>BMI</th>
<th>IPAP</th>
<th>EPAP</th>
<th>Back up rate</th>
<th>Daily hours</th>
<th>Ventilation (yrs)</th>
<th>Airway Clearance</th>
<th>Compliant with LVR or MIE</th>
<th>Size of initial pneumothorax (cm)</th>
<th>Diagnostic modality</th>
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<td>M</td>
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<td>65</td>
<td>19</td>
<td>22</td>
<td>10</td>
<td>16</td>
<td>9</td>
<td>0.25</td>
<td>LVR</td>
<td>Yes</td>
<td>12</td>
<td>CXR AP</td>
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<tr>
<td>2</td>
<td>24</td>
<td>F</td>
<td>Limb Girdle Dystrophy</td>
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<td>50</td>
<td>19</td>
<td>20</td>
<td>4</td>
<td>20</td>
<td>8</td>
<td>4</td>
<td>LVR</td>
<td>No</td>
<td>&lt;0.5</td>
<td>CXR PA</td>
</tr>
<tr>
<td>3</td>
<td>53</td>
<td>F</td>
<td>CAP myopathy</td>
<td>0.6</td>
<td>42</td>
<td>16</td>
<td>24</td>
<td>5</td>
<td>16</td>
<td>22</td>
<td>2</td>
<td>LVR</td>
<td>No</td>
<td>&lt;1</td>
<td>CT</td>
</tr>
<tr>
<td>4</td>
<td>26</td>
<td>F</td>
<td>Collagen 6 disease</td>
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<td>27</td>
<td>13</td>
<td>20</td>
<td>4</td>
<td>20</td>
<td>8</td>
<td>6.5</td>
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<td>Yes (bid)</td>
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<td>CT</td>
</tr>
<tr>
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<td>F</td>
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<td>16</td>
<td>6</td>
<td>14</td>
<td>15</td>
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<td>MIE + LVR</td>
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<td>2.5</td>
<td>CXR PA, CT</td>
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<td>Kyphoscoliosis and Ankylosing spondylitis</td>
<td>0.83</td>
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<td>14</td>
<td>20</td>
<td>6</td>
<td>20</td>
<td>9.5</td>
<td>9</td>
<td>LVR</td>
<td>Yes (with adhesions)</td>
<td>large with</td>
<td>CXR PA</td>
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</table>

**Figure 1B** Follow up CT of ALS patient with residual blebs at right lung base

**Figure 2A** Radiograph demonstrating the sentinel event in patient with limb girdle muscular dystrophy, a left apical pneumothorax measuring <1 cm

**Figure 2B** Large right pneumothorax in a patient with limb girdle muscular dystrophy, presenting 2 months after initial left pneumothorax

**Figure 2C** One-week post doxycycline pleurodesis of the right pleura in patient with limb girdle muscular dystrophy
Pneumothorax in some chest effected patients

Case 4
A 24-year-old female with collagen 6 disease who had had a history of scoliosis and Harrington rod placement with multiple revisions. The patient was given a single trial of mouthpiece ventilation for persistent hypercapnia despite nocturnal ventilation and daytime dyspnea. Settings on a Legendaire ventilator were VT 600 mL, PEEP 0 cm H2O, waveform sine, rate of 5 breaths per minute and I time of 25%. Pressure alarms were set at minimum 2 cm H2O and maximum 38 cm H2O. The patient experienced right-sided chest pain later in the evening and night after the mouthpiece ventilation trial. Upon presentation to the emergency department she had a small 1.3 cm right sided pneumothorax (Figure 4). An 8 French chest tube was initially used and then replaced with an 18 French chest tube due to persistent air leak. NIV settings were maintained. After eventual re-expansion of the lung and clamping of the chest tube it was withdrawn, however pneumothorax recurred within 6 hours, necessitating a third tube. The total duration of chest tube therapy was 62 days. The patient went on to use bi-level non-invasive positive airway pressure ventilation increasingly through the day and night time and did not re-trial mouthpiece ventilation subsequently.

Case 5
A 55-year-old female with myotonic dystrophy type 1 had a history of recurrent aspiration and interstitial lung changes related to this. The patient had declined enteral nutrition with gastrostomy and so continued to maintain oral intake despite known aspiration risks. The patient was using nocturnal NIV with LVR airway clearance technique. The patient presented with a large right pneumothorax (Figure 5A) which was treated with a 10 French chest tube, with resolution and removal of chest tube in 16 days. During this time MI-E was held. The patient did experience a recurrence 7 months later after resuming her usual NIV settings, LVR and mouthpiece ventilation (Figure 5B). At this time both chemical and surgical pleurodesis were considered, but declined due to the patient’s overall condition.

Case 6
A 64-year-old female had ankylosing spondylitis and kyphoscoliosis with Harrington rod placement. She had no known interstitial lung disease. This patient was using nocturnal NIV with daytime LVR airway clearance technique. The patient presented with a large pneumothorax (Figure 6A) which was treated with a 10 French chest tube, with resolution and removal of chest tube in 16 days. During this time MI-E was held. The patient did experience a recurrence 7 months later after resuming her usual NIV settings, LVR and mouthpiece ventilation (Figure 6B). At this time both chemical and surgical pleurodesis were considered, but declined due to the patient’s overall condition.
It is interesting that pneumothorax was observed in these stable neuromuscular patients in whom lung compliance is assumed to be normal. Low BMI was seen in this patient group. Malnutrition has been associated with spontaneous pneumothorax in patients with anorexia nervosa (19). Decreased total lung protein may play a role in the pathophysiology of pneumothorax in a state of malnutrition as described in an animal model (20) and therefore may play a significant role in the development of pneumothorax in neuromuscular patients.

CONCLUSION

The management of pneumothorax in neuromuscular patients dependent on chronic non-invasive ventilation requires special consideration. Removing positive airway pressure ventilator support is not an option, and this is perhaps why early chest tube management and prolonged chest tube duration was seen in this group. Lung volume recruitment maneuvers or MIE were suspended until pneumothorax was resolved, but with careful monitoring to ensure de-recruitment and retained airway secretions did not further compromise the patient. These maneuvers were typically restarted in 46 weeks following the pneumothorax event.

Talc pleurodesis has associated with an ARDS-type reaction, dyspnea and increased oxygen requirements in patients, although studies have been mixed as to the incidence of these side-effects (21,22). In patients with chronic respiratory insufficiency and NIV dependence, it is important to consider the risk of precipitating acute respiratory failure with pleurodesis. One patient in this series did undergo successful chemical pleurodesis with doxycycline in the ICU, without adverse sequelae. This patient did not experience recurrence on this side.

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REFERENCES