HHHFNC with oxygen therapy as treatment for pneumothorax in a patient with spinal muscular atrophy type I

Ronald Cheuk-Man Fung and Ada Yuen-Fong Yip

1Department of Paediatrics and Adolescent Medicine, Princess Margaret Hospital; 2Department of Paediatrics, Kwong Wah Hospital, Hong Kong

Abstract
We reported a 19-year-old bilevel positive pressure (BiPAP) dependent spinal muscular atrophy (SMA) patient, who developed recurrent pneumothorax and was managed successfully with heated, humidified high flow nasal cannula (HHHFNC) with oxygen therapy.

Keywords: Heated humidified high flow nasal cannula, HHHFNC, pneumothorax, spinal muscular atrophy

Case report
We report a 19-year-old bilevel positive pressure (BiPAP) dependent spinal muscular atrophy (SMA) patient, who developed recurrent pneumothorax and was managed successfully with heated, humidified high flow nasal cannula (HHHFNC) with oxygen therapy.

Our patient was diagnosed to have suffered from SMA type I before 6 months of age. As expected from the natural course, she gradually developed restrictive lung disease, kyphoscoliosis and obstructive sleep apnoea. Despite regular chest physiotherapy and mucous clearance program for restrictive lung disease, prompt treatment for any chest infection, spinal surgery for kyphoscoliosis and nocturnal biphasic positive airway pressure (BiPAP) for obstructive sleep apnoea, she developed chronic respiratory failure with a restrictive lung function. FEV$_1$ was 21% predicted, FVC 20% predicted, FEV$_1$/FVC 108% predicted. Her respiratory muscle strength was diminished with maximum inspiratory pressure being -43 cmH$_2$O and maximum expiratory pressure being +25 cmH$_2$O. She was BiPAP dependent for at least 12 hours per day.

The patient tolerated BiPAP (ST with AVAPS mode, VT 370 ml (8.5 ml/kg), PIP 15-18 cmH$_2$O, PEEP 8 cmH$_2$O, rate 14/min, I:E ratio 1:6) well until 16 months ago when she developed 4 episodes of pneumothorax (Table 1). All episodes of pneumothorax were small in size and were on the left side. Chest pain was the only presenting symptom and she remained haemodynamically stable with no respiratory distress, desaturation nor carbon dioxide retention. No chest drain insertion was required. She could not tolerate BiPAP even with a lower pressure limit settings nor continuous positive airway pressure support because of increased chest pain. Hence, HHHFNC provided by Airvo (Fisher-Paykel) supplemented with oxygen was given to provide ventilation support and this was much better tolerated. We titrated the flow rate initially against respiratory rate, oxygen saturation, blood gases and comfort level. Optimal flow rate was found to be 35 L/min with 7 L/min oxygen, i.e. FiO$_2$ 0.37, was given via nasal cannula. Sleep polysomnography was done that showed obstructive hypopnoea index being 3.5 and central apnea index of 1.2. There was no significant desaturation and TcCO$_2$ was normal throughout the night.

In view of recurrent pneumothoraxes, computed tomography thorax (Figures 1-3) was done. It showed left apical bullae and bronchiectatic changes at left upper and lower lobes. Chemical pleurodesis was declined by patient. Placement of valve prosthesis, with an aim to collapse the apical lung so as to prevent further pneumothorax and to reduce ventilation-perfusion mismatch due to bronchiectasis, was deemed not suitable in view of her poor lung function reserve and the absence of continued air leak that is necessary to confirm correct placement of valve. Bullectomy with mechanical pleurodesis was also deemed too high risk for the poor lung function.

*Author to whom correspondence should be addressed.
Email: yyfz02@ha.org.hk
### Table 1. Details of recurrent pneumothoraxes

<table>
<thead>
<tr>
<th>Episode</th>
<th>Time</th>
<th>Respiratory support</th>
<th>Duration of pneumothorax</th>
<th>Blood gas</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>August 2012</td>
<td>Optiflow 10 L/min</td>
<td>3 days</td>
<td>pH 7.45, pCO(_2) 4.2 kPa, pO(_2) 16.1 kPa, HCO(_3) 21.3 mmol/L, BE -1.8</td>
</tr>
<tr>
<td>2</td>
<td>October 2013</td>
<td>AIRVO 20 L/min</td>
<td>10 days</td>
<td>pH 7.45, pCO(_2) 4 kPa, pO(_2) 15 kPa, HCO(_3) 20.7 mmol/L, BE -2</td>
</tr>
<tr>
<td>3</td>
<td>November 2013</td>
<td>AIRVO 20 L/min</td>
<td>3 days</td>
<td>pH 7.4, pCO(_2) 4.5 kPa, pO(_2) 12.8 kPa, HCO(_3) 20.4 mmol/L, BE -3.6</td>
</tr>
<tr>
<td>4</td>
<td>December 2013</td>
<td>AIRVO 36 L/min</td>
<td>21 days</td>
<td>pH 7.42, pCO(_2) 5.2 kPa, pO(_2) 8.6 kPa, HCO(_3) 24.8 mmol/L, BE 0.5</td>
</tr>
</tbody>
</table>

---

**Figure 1.** Bronchiectatic changes at left lower lob.

**Figure 2.** Multiple bullae at left apical lung.
Discussion

The development of pneumothorax in a non-invasive ventilator dependent patient, like our SMA patient, can be a challenging clinical situation. We tackled the problem and kept our patient adequately ventilated by offering HHHFNC supplemented with oxygen therapy.

Recurrent pneumothoraces

Literature search did not find any information concerning significant association between SMA and pneumothorax. The recurrent episodes of air leak in our patient probably result from multiple predisposing factors like kyphoscoliosis, repeated pneumonia leading to bronchiectasis and lung cysts and prolonged BiPAP ventilation. Neuromuscular patients are prone to have micro-atelectasis secondary to hypoventilation. Kyphoscoliosis causes areas of lung collapse and hyperinflation due to ball valve effect. Mucous clearance was impaired and result in chest infection. Recurrent infection causes damage to lung parenchyma and hence, development of bronchiectasis. With prolong use of positive pressure ventilation, these patients are prone to have lung cyst formation and pneumothorax. Needless to say, BiPAP use itself is also a risk factor for pneumothorax.4

Use of HHHFNC with oxygen Therapy in pneumothorax

Similar to ordinary high flow oxygen therapy, HHHFNC with oxygen therapy can hasten resolution of pneumothorax by lowering the partial pressure of nitrogen in pleural cavity. It is more comfortable and is generally better tolerated by patients as the heated and humidified air flow prevents upper airway mucosa from drying up. In addition, HHHFNC with oxygen therapy provides ventilatory support to patient who suffers from chronic respiratory failure. The exact mechanism on how HHHFNC with oxygen therapy works is not fully understood but it has been shown to work by decreasing the work of breathing and decreasing energy spend on inspired gas conditioning.

The use of high flow therapy decreases the work of breathing by (1) washing out dead-space in pharynx so as to improve alveolar minute ventilation; (2) providing gas flow that is higher than spontaneous inspiratory flow so that inspiratory resistance at nasopharyngeal level is minimised; (3) providing distending pressure to the lung and improves alveolar recruitment. The administration of heated and humidified gases has been demonstrated to be able to (4) improve airway conductance and lung compliance, hence decreases the work of breathing as well. In addition, it (5) reduces energy required for conditioning inspiratory gas and hence decreases the oxygen consumption.

Setting of HHHFNC

HHHFNC therapy can deliver continuous high flow ranged from 0.3 to 60 L/min with 100% relative humidity (that is about 44 mgH2O/L) at 37°C. To achieve this, the humidifier has to set at temperature ≥37°C and an appropriate sized nasal cannula has to be chosen. Nasal cannula should not be too large that lead to inadvertent high pressure nor to small that lead to excessive air leak around nostrils. For the Fisher-Paykel system, the infant sized nasal cannula size provides maximum flow rate up to 7 L/min, paediatric size up to 8 L/min, adult size up to 50 L/min. Various oxygen concentrations can be delivered with the incorporation of air-oxygen blender.

Flow rate should be titrated against patient’s respiratory rate, work of breathing, oxygen saturation and arterial blood gas. Till now, there is no consensus as to how it should be achieved. Some groups recommend giving 100% oxygen and titrate the flow rate up from 0 L/min with a 0.5-1 L/min increment until the desired effect is observed, then wean the oxygen supplement to minimum required level. However, we followed a different approach in view of her poor lung functions and started from a flow rate equivalent to 2-3 times of

Figure 3. Small left apical pneumothorax.
patient's minute ventilation. We believe that this is a more logical approach as this formula gives a good estimation of flow rate required to exceed that of inspiration, so that pharyngeal dead space is washed out and nasopharyngeal resistance is decreased.

This report highlights the dilemma of managing pneumothorax in a BiPAP dependent patient. It also demonstrates the success of HHHFNC with oxygen therapy in replacing BiPAP as a means of ventilation for patient with chronic respiratory failure. More research is needed in the use of HHHFNC in management of obstructive sleep apnoea complicated with pneumomthorax.

References