LETTER

Survey of the use of non-invasive positive pressure ventilation in UK and Australasian children with cystic fibrosis

Non-invasive positive pressure ventilation (NIPPV) for respiratory failure in cystic fibrosis (CF) is frequently used in adults and has been shown to be of benefit to patients with advanced disease in terms of stabilisation of lung function, reduction in symptoms and increased exercise capacity. When used as an adjunct to physiotherapy, NIPPV increases oxygen saturations (SpO2), tidal volume, maximum expiratory muscle strength and ease of sputum clearance.34 There are no guidelines for the assessment of gas exchange or timing and mode of NIPPV initiation in patients with CF. The British Thoracic Society guideline states there is insufficient evidence to recommend its routine use in patients with CF.5 The aim of this study was to establish current practice with regard to investigation of respiratory failure, factors leading to NIPPV initiation and extent of the use of this modality across UK and Australasian (ANZ) paediatric CF centres.

A semi-structured questionnaire consisting of 21 closed and open-ended questions was sent to the lead CF consultant and CF physiotherapists of specialist paediatric CF centres in the UK (n=27) and ANZ (n=14). The response rate was 82% (25 UK centres, 11 ANZ centres), representing a total of 5954 children. Twenty-three children (0.39%) from 13 centres were using NIPPV (11 UK and 12 ANZ). The median (range) age of NIPPV initiation was 14 (6–17) years and the median (range) usage of NIPPV per night was 8 (3–10) h. Eleven of the 36 centres (31%) reported that they have a protocol for NIPPV initiation, but it was unclear whether this was specific for CF. The preferred mode of NIPPV was bi-level NIPPV (75%), followed by single-level preset pressure ventilation (19%), volume control single-level ventilation (3%) and continuous positive airways pressure (3%). Nasal masks were the most frequently used interface (47%), followed by full face masks (38%), mouthpieces (13%) and nasal pillows (2%).

Less than half of the CF centres (17/36) undertook CF sleep studies. SpO2 monitoring alone was most commonly used (51% of centres), followed by SpO2 and transcutaneous carbon dioxide monitoring (22%). Full polysomnography was less frequently used as a first-line investigation (16.2%). Assessment of respiratory failure differed between childhood CF centres, with different definitions for hypoxia and hypercapnia in use (table 1).

The principal reasons for initiating NIPPV included an acute exacerbation, as a bridge to transplant and as an adjunct to physiotherapy. There were 19 reported NIPPV failures in children with CF from 10 centres. Reasons for failure included claustrophobia, inability to tolerate pressure, discomfort, poor initial set-up, parent or patient anxiety and poor adherence. Four adverse events were reported (issues with mask fitting and pressure sores, n=2; retained seclusions, n=1; and pneumothorax, n=1).

NIPPV is rarely used in UK and ANZ paediatric CF populations, probably due to improved patient outcomes and the very low prevalence of respiratory failure in childhood CF. Bi-level NIPPV is the preferred mode of ventilation. However, there is no agreed definition of hypoxia and hypercapnia, no uniformity in assessing gas exchange and no standard protocol for the indications and institution of NIPPV in children with CF. As very few of the expected benefits of NIPPV have been proven, particularly in the paediatric CF population, and as there is a high frequency of NIPPV failure, the need for further research in this area is highlighted, beginning with the need for protocols to be developed and evaluated.

Table 1 Variation in hypoxia and hypercapnia definition used between CF centres in the UK and Australasia (16 centres were unsure of the definitions)

<table>
<thead>
<tr>
<th>Hypoxia</th>
<th>Hypercapnia</th>
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<tbody>
<tr>
<td>Frequent desaturations &lt;90%</td>
<td>PetCO2 &gt;6.7 kPa</td>
</tr>
<tr>
<td>Baseline SaO2 &lt;93%</td>
<td>CO2 &gt;6.7 kPa for &gt;25% sleep study</td>
</tr>
<tr>
<td>SpO2 &lt;90% for &gt;10% sleep study</td>
<td>TcCO2 rise to 6.7 kPa or rise by 0.9 kPa during sleep</td>
</tr>
<tr>
<td>SpO2 &lt;92% for &gt;5% sleep study</td>
<td>CO2 &gt;7 kPa</td>
</tr>
</tbody>
</table>

CO2, carbon dioxide; PetCO2, end tidal carbon dioxide pressure; SaO2, arterial oxygen saturations; SpO2, non-invasive oxygen saturations; TcCO2, transcutaneous carbon dioxide pressure.
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