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. With advanced disease in terms of stabilisation of lung function, reduction in symptoms and increased exercise capacity, NIPPV is frequently used as a bridge to transplantation and as an adjunct to physiotherapy.

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’. 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 15 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 polysomography 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 secretions, 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.

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>
</tr>
</thead>
<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>TCO₂ 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; Saa₂, arterial oxygen saturations; SpO₂, non-invasive oxygen saturations; TCO₂, transcutaneous carbon dioxide pressure.

Acknowledgements The authors acknowledge Dr Michelle Chatwin and Dr Ian Balfour-Lynn, Royal Brompton Hospital, London, UK. N Collins, G Gupta, S Wright, L Gauld, D Urquhart, A Bush. Department of Physiotherapy, Royal Brompton Hospital, London, UK; Department of Paediatric Respiratory Medicine Royal Brompton Hospital and Imperial College, London, UK; Royal Brisbane Children’s Hospital, Brisbane, Australia; Mater Children’s Hospital, Brisbane, Australia.

Correspondence to Nicola Collins, Physiotherapy Department, Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK; n.collins@rbht.nhs.uk

Competing interests None.

Provenance and peer review Not commissioned; externally peer reviewed.

Accepted 19 May 2010

Thorax 2010;65:1–11

doi:10.1136/thx.2010.139063

REFERENCES

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

N Collins, A Gupta, S Wright, L Gauld, D Urquhart and A Bush

Thorax  published online September 3, 2010

Updated information and services can be found at:
http://thorax.bmj.com/content/early/2010/09/03/thx.2010.139063

These include:

References
This article cites 5 articles, 3 of which you can access for free at:
http://thorax.bmj.com/content/early/2010/09/03/thx.2010.139063#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/