



# British Thoracic Society guideline for respiratory management of children with neuromuscular weakness: commentary

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## ABSTRACT

The British Thoracic Society guideline for respiratory management of children with neuromuscular weakness summarises the available evidence in this field and provides recommendations that will aid healthcare professionals in delivering good quality patient care.

Neuromuscular diseases as a group are relatively common with a prevalence of about 1 in 3000. Neuromuscular weakness (NMW) can directly reduce respiratory muscle strength and compromise upper airway muscle tone, airway protection and spinal support. The respiratory consequences are hypoventilation, upper airway obstruction, aspiration lung disease, secretion retention and lower airway infection, and the mechanical effects of progressive scoliosis. Acute respiratory failure associated with respiratory infection is the most frequent reason for unplanned hospital admission, and chronic respiratory failure is a frequent cause of death. With appropriate intervention, the incidence of unplanned hospital admission can be reduced and life expectancy can be improved.

This British Thoracic Society guideline<sup>1</sup> attempts to summarise the available evidence in this field and provides recommendations that will aid healthcare professionals in delivering good quality patient care. Many of the principles of respiratory management are not disease specific and the objective of this guideline is to provide recommendations that can be applied to all children with NMW. The evidence for much of current practice is weak and is based largely on observational studies. The Guideline Committee attempted to identify and summarise the existing evidence, and when that is lacking, provide expert consensus opinion. The guideline starts with a background overview of respiratory problems in children with NMW. A brief summary of the conditions covered by the guideline is provided in an appendix. The respiratory management of children with NMW is then covered in eight sections:

- ▶ Identifying children at risk of respiratory complications
- ▶ Airway clearance and respiratory muscle training
- ▶ Assisted ventilation
- ▶ Planning for surgical procedures
- ▶ Scoliosis
- ▶ Other interventions that impact on respiratory health
- ▶ Transition to adult care
- ▶ Quality of life and palliative care

A summary of the recommendations and good practice points for each of these sections is provided at the beginning of the guideline. In this commentary, I am going to focus on two aspects of the respiratory care of children with NMW which cause particular anxiety for clinical teams, carers and families: the management of acute respiratory failure and the identification and management of chronic hypoventilation.

## ACUTE RESPIRATORY FAILURE

In children with NMW, otherwise minor viral respiratory tract infections can result in increased airway mucus production and reduced respiratory muscle strength, a combination which can lead to atelectasis, worsening lung compliance, increased respiratory work and eventually respiratory failure. The guideline recommends that clinical assessment of respiratory health should be part of every medical consultation for children with NMW and this should include an assessment of cough effectiveness.

The most appropriate and effective methods of secretion clearance will vary from child to child. Cough augmentation, which may be necessary, involves two steps—first to increase inspiratory volumes, and second to increase the expiratory forces applied to the inspired volume. Simple methods should be considered first; for example, use of breath-stacking or lung volume recruitment bags, combined with manual compression of the chest and abdomen. Although mechanical insufflation/exsufflation (MI-E) devices can be very effective, there are as yet no studies which show that they are superior to other methods of assisting cough.

Pulse oximetry at home and in the hospital can provide a useful trigger for the early institution of aggressive airway clearance during respiratory tract infection. Children with NMW who are experiencing a respiratory exacerbation and who have oxygen saturation below 95% in room air despite airway clearance carried out by their carers or parents at home will require admission to a hospital unit familiar with the management of this problem. Experienced practitioners (usually physiotherapists) should institute immediate and regular airway clearance methods, which may be needed several times a day for several days. The ability to use a range of different airway clearance techniques, including MI-E devices, is important.

In the event that a child requires respiratory support, non-invasive ventilation (NIV) is often

successful and the guideline recommends that it should be used as first-line treatment for acute respiratory failure. If intubation and ventilation are required, subsequent extubation is more likely to be successful if it is delayed until airway secretions are minimal, supplemental oxygen has not been required for more than 12 h, and if effective airway clearance methods are used before and after extubation.

### CHRONIC HYPOVENTILATION

In progressive neuromuscular disease the first indication of respiratory failure is usually sleep-associated hypoventilation. Symptoms of hypoventilation at night are non-specific, often come on gradually and may go unnoticed for several months. Children who hypoventilate at night are at increased risk of acute respiratory failure during respiratory infection. NIV is effective at reversing the symptoms of hypoventilation and reducing the frequency of unplanned hospital admissions.

The likelihood of hypoventilation correlates with the severity of weakness of the respiratory muscles, and this can be assessed in several ways; vital capacity is one of the easiest to measure and for Duchenne muscular dystrophy, of the available tests of respiratory muscle strength, it is the best predictor of nocturnal hypoventilation, albeit with rather poor sensitivity and specificity.

When hypoventilation is suspected further assessment will require some form of overnight monitoring. Good quality oximetry is sufficient in most children to exclude clinically significant hypoventilation. When oximetry is abnormal, more detailed recordings are recommended to elucidate the nature of the problem.

NIV is recommended for symptomatic nocturnal hypoventilation and for all children with daytime hypoventilation. NIV during sleep may correct daytime hypercapnia, probably by re-setting chemoreceptor sensitivity to carbon dioxide. In children with more severe weakness, respiratory failure with hypercapnia can occur during wakefulness and during sleep to an extent that is not reversed by nocturnal assisted ventilation alone. For these children daytime NIV is used to relieve

symptoms of hypoventilation, improve quality of life and increase life expectancy. When children require ventilation for more than 16 h in each 24 h period, a tracheostomy may provide a more satisfactory interface between the child and the ventilator than a mask. Tracheostomy may also be necessary in children with severe bulbar dysfunction. The decision to insert a tracheostomy is often difficult and the guideline recommends that the preferences of the family and child or young person, and the experience of the clinical team, need to be taken into account. Once children have been established on NIV they need to be monitored regularly. The guideline recommends a minimum of overnight oxycapnography once every 12 months in stable children. Repeat assessment should be more frequent in infants, and in children who have symptoms of hypoventilation or disturbed sleep, or who have troublesome respiratory exacerbations.

The guidelines do not specify who should care for children with respiratory problems associated with NMW. The recommendations require expertise in respiratory assessments including interpretation of sleep monitoring data, availability of physiotherapists experienced in the use of appropriate airway clearance methods, including MI-E devices, clinical nurse specialists or physiotherapists expert in providing support to children and their families when NIV is needed, and access to intensive care units with experience in managing children with NMW. For most children this will mean attendance at a tertiary respiratory centre for at least some of their care. Close liaison between the tertiary centre and local hospital units is essential. It is hoped that the guidelines will provide a framework for good practice for all those involved in the respiratory care of children with NMW.

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### REFERENCE

1. Hull J, Anupravan R, Chan E, *et al*. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. *Thorax* 2012;**67** (Suppl 1):1–40.