Workshop report

Recommendations for respiratory care of adults with muscle disorders

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The Muscular Dystrophy Campaign convened a workshop on 2nd February 2005 to draw up evidence-based recommendations for the management of respiratory problems in adults with muscle diseases. The strength of these recommendations was graded using the Scottish Intercollegiate Guidelines Network (SIGN) methodology (Table 1). Neurogenic conditions, neuromuscular junction disorders and myotonic dystrophies were excluded, although the principles of practice recommended will apply to many of these conditions.

1. Clinical assessment

- Assess respiratory function annually in patients with muscle diseases in which respiratory muscle involvement is known to occur inevitably or frequently (Table 2). [D]
- See patients with a low vital capacity (VC) – less than 1.5 litres – or with rapidly progressive disease more frequently. [D]
- Check for symptoms (Table 3) and signs (Table 4) of respiratory failure at each clinic visit. [D]
- Use VC to monitor respiratory function, where practical with sitting and supine measurements. [D]
- Use maximum mouth pressures and sniff nasal inspiratory pressure to detect early respiratory muscle weakness. They are not needed routinely to monitor respiratory function, since they can be tiring for patients and are not essential once the presence of respiratory muscle weakness has been established. [D]
- Measure peak cough flow (PCF) to monitor expiratory function. [D]
- Measure daytime oxygen saturation (SpO2) in patients with respiratory symptoms or signs, and in those with a VC < 1.5 litres. [D]
- Measure arterial blood gases if daytime SpO2 is less than 94%, if signs or symptoms suggest respiratory failure, or if there are symptoms of nocturnal hypoventilation. [D]
- Perform sleep studies annually when VC is less than 1.5 litres, or if there are symptoms of nocturnal hypoventilation. [D]

2. Prophylaxis

- Advise pneumococcal vaccination and annual influenza vaccination to patients with a VC < 50% predicted. [B]
- Offer preconception counselling to female patients of childbearing age about the impact of pregnancy on their respiratory function. [D]
- Give patients with respiratory muscle weakness a prescription for antibiotics to keep at home. Advise them to have a low threshold for commencing antibiotics when they develop a respiratory tract infection. [D]
- Offer respiratory muscle training to motivated patients with mild or moderate respiratory muscle weakness. [C]
- There is no evidence to support “prophylactic” nocturnal non-invasive ventilation (NIV) in the absence of daytime respiratory failure or symptoms of nocturnal hypoventilation. [D]
3. Specialist referral

- Refer patients who exhibit symptoms and signs of possible bulbar dysfunction to a speech and language therapist for advice on reducing the risks of aspiration. [D]

- Refer patients with a weak cough, in whom the PCF is usually less than 270 l/min, to a respiratory physiotherapist for education in assisted cough techniques. [D]

- Exclude other cardiac (Table 5) and respiratory problems before attributing breathlessness to the underlying muscle condition. [D]

4. Non-invasive ventilation

- Do not use oxygen to treat hypercapnic respiratory failure, except in conjunction with NIV or in palliative care. [C]
- Offer NIV to patients with diurnal ventilatory failure or symptomatic sleep disordered breathing. [C]
- Consider invasive ventilation in an intensive care unit (ICU) if the patient is too unwell to be safely managed with NIV. [D]
- Offer invasive ventilation through a tracheostomy as an alternative to NIV in selected cases.
- Consult a specialist assisted ventilation service before a tracheostomy is performed on ICU. [D]

5. Care pathways

- Draw up an optimal management plan for illnesses unrelated to the muscle condition, and discuss with a specialist neuromuscular specialist team how it can be best delivered. This also applies to patients who require invasive investigations or operative procedures. [D]
- Provide patients with information on possible treatment options available should they develop respiratory failure. [D]
- Encourage the preparation of advance directives. [D]
- Manage patients in liaison with a specialist unit which has access to neurology, respiratory, rehabilitation medicine, speech and language therapists, physiotherapists, dietitians and any other relevant specialists. [D]
- Make arrangements to ensure that the transition from childhood to adulthood is smooth. [D]
- Provide access to a unit which runs a 24 h NIV service and has specialist respiratory physiotherapy, in the event of an acute respiratory crisis. [D]

6. Workshop participants

Dr. Robert Bullock, Consultant Anaesthetist, Newcastle General Hospital, Newcastle-Upon-Tyne.
Dr. Brendan Cooper, Consultant Clinical Scientist, Lung Investigation Unit, University Hospital, Birmingham.
Dr. Michelle Eagle, Physiotherapy Research Practitioner in NMD, Muscle Centre, Newcastle.
Dr. David Hilton-Jones, Consultant Neurologist, Radcliffe Infirmary, Oxford.
Dr. Robin Howard, Consultant Neurologist, National Hospital for Neurology and Neurosurgery, Queen’s Square, London.
Dr. William Kinnear, Consultant in Respiratory Medicine, Queens Medical Centre, Nottingham (Chair).
Dr. Margaret Phillips, Consultant in Rehabilitation Medicine, Division of Rehabilitation and Ageing, University of Nottingham, Derby City Hospital, Derby.
Dr. Tina Shahrizaila, Specialist Registrar in Neurology, Queens Medical Centre, Nottingham (Secretary).
Dr. Anita Simonds, Consultant in Respiratory Medicine, Royal Brompton Hospital, London.

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