

6.8 LONG TERM VENTILATION IN CHILDREN AND YOUNG PEOPLE

Some conditions are such that a child is unable to sustain adequate breathing or ventilation on their own and they need assistance or long term ventilation (LTV).

Children and young people who have experienced a traumatic spinal cord injury may never be able to breathe spontaneously and require LTV twenty fours each day. More commonly a group of children and young people who have progressive neuromuscular disorders, such as duchenne muscular dystrophy (DMD) or spinal muscular atrophy (SMA), require LTV to treat nocturnal hypoventilation (breathing which is inadequate while asleep) known as nocturnal non invasive ventilation (NIV). These children can breathe adequately during the day while awake, but the timely introduction of NNIV can rest the respiratory muscles at night and greatly improve day time quality of life. NIV is often beneficial before scoliosis surgery in children with DMD or SMA.

Children with weak respiratory muscles will also have weak coughing and be unable to clear secretions especially when suffering a respiratory infection. Modern physiotherapy with 'cough assist' techniques and devices is an equally important management tool.

LTV has an established track record in patients with ventilatory failure. There are currently 27 children on LTV support in N Ireland. The numbers are likely to increase as the benefits of this treatment are realised for children with neuromuscular disease (NMD). Respiratory insufficiency is the most common cause of early death in children and adolescents with NMD. In the last decade nocturnal non invasive ventilation (NNIV) has considerably extended survival in children with these conditions and simultaneously vastly improved quality of life.

Standards of care have been drawn up for both DMD and SMA. . These complex conditions require a multidisciplinary team approach involving a number of different specialties. These should be available at a one stop shop multidisciplinary outpatient clinic. which should be based at the Royal Belfast Hospital for Sick Children, but delivered at the child's home.

Overarching standard 33:

Assessment and support

All children and young people and young people requiring or potentially requiring long term ventilation (LTV) or nocturnal non invasive ventilatory (NNIV) support at home should have access to a specialist multidisciplinary team at tertiary level.

Rationale:

LTV has an established track record in patients with ventilatory failure (such as children with neuromuscular diseases). In some LTV involves 24 hour respiratory support (e.g. spinal cord injuries), while in the majority the goal is to treat nocturnal hypoventilation and thereby improve day time quality of life. Nocturnal non invasive ventilation (NIV) is often beneficial before scoliosis surgery in children with duchenne muscular dystrophy and spinal muscular atrophy.

Successful home management of ventilator dependent children can be traced to a smooth collaborative discharge from hospital to home and, once at home, a streamlined patient centred process with direct lines of communication.

Evidence:

Chest 2006; 130: 1879-1886. Recent advances in respiratory care for neuromuscular disease.

<http://www.chestjournal.org/cgi/content/full/130/6/1879>

Clinical Respiratory Journal 2008. (DOI: 10.1111/j.1752-699X.2007.00034.x) Impact of home mechanical ventilation on health-related quality of life in patients with chronic alveolar hypoventilation: a prospective study.

<http://www3.interscience.wiley.com/cgi-bin/fulltext/119423626/PDFSTART>

Chest 2007; 132: 1977-1986. American College of Chest Physicians Consensus Statement on the respiratory and related management of patients with Duchenne Muscular Dystrophy undergoing anaesthesia or sedation

<http://www.ncbi.nlm.nih.gov/pubmed/18079231?dopt=Abstract>

J child Neurology. 2007; 22(8); 1027-1049. Consensus statement for standard of care in spinal muscular atrophy. <http://intl-icn.sagepub.com/cgi/reprint/22/8/1027>

Responsibility for delivery / implementation			
HSC Trusts			
Quality Dimension			
<p>1. Children and young people potentially or requiring LTV should have access to:</p> <ul style="list-style-type: none"> • physical and psychosocial assessment by a specialist multidisciplinary team; • appropriate equipment within 4 weeks (with back up technical and equipment support); • a clearly identified link respiratory nurse; and, • trained carers within 6 months from being clinically suitable for discharge <p>2. Children with neuromuscular disease requiring LTV or NNIV should have access to a physiotherapist trained to meet their needs and familiar with cough assist techniques and devices.</p>			
Performance Indicator	Data source	Anticipated Performance Level	Date to be achieved by
Percentage of children and young people requiring NNIV/LTV who receive an initial assessment/management (within 4 weeks) and regular follow up (at least twice yearly) by the specialist regional multidisciplinary team	Belfast HSC Trust Regional database Audit	50%	March 2011
Percentage of children and young people with a named link specialist respiratory nurse for direct access to the regional multidisciplinary team	Belfast HSC Trust Patient Satisfaction Survey	70% 90%	March 2011 March 2012