Section 1: Introduction

Reshma Amin, Ian MacLusky & David Zielinski on behalf of the CTS Pediatric Home Ventilation Guidelines Panel

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Section 1: Introduction

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Introduction

Coincident with the move to home mechanical ventilation (HMV) for adults, HMV of children has also become a standard of practice for a variety of conditions that result in chronic respiratory failure. There are similar driving forces for both children and adults that explain the rising incidence of HMV, such as rising costs of hospital care and the effort to improve quality of life (QoL) and normal development. However, some factors are specific for children, such as the advent of commercially available non-invasive masks and positive pressure ventilators adapted for use in children. As a consequence, HMV for children has, over the last 30 years, evolved from being an infrequently offered option to standard of care, with increasing numbers of children receiving this treatment worldwide. Despite this, there remain many areas in which there are little research data to guide clinical practice, resulting in neither consistency nor agreed-upon protocols. This guideline document is an attempt to provide answers to several key questions:

1. Which patients would most benefit from long-term HMV, and can we quantify and predict these benefits? How can we identify the optimum methodology (invasive versus non-invasive) in specific diseases and children?
2. What are the optimal timing and indications for initiation of ventilation (invasive and non-invasive)? What criteria should be used for the timing of initiation? Is there any therapeutic benefit for early initiation (i.e., prior to the development of overt hypoventilation)?
3. What are the optimal requirements for ongoing monitoring and follow-up (addressing changes in clinical condition), both at home and within the healthcare system?
4. What are the ideal (or even minimum) required supports for the family, not only technological, but also financial and personnel, to allow for a child to be successfully cared for at home on long-term ventilation (invasive or non-invasive)?

Objective

These guidelines are designed as a complementary document to the adult guidelines already published.\textsuperscript{1} The objective of the present clinical practice guideline is to provide guidance to pediatric patients, their families and caregivers and healthcare teams on the role of HMV for a variety of patient populations, as well as common issues around its initiation and long-term maintenance in the community. Specifically, this guideline focuses on invasive ventilation and positive pressure non-invasive ventilation (NIV). Continuous positive pressure ventilation (CPAP) is not discussed. Additionally, negative pressure and positive pressure body ventilators have been excluded from these guidelines, as they have been replaced with portable positive pressure ventilators.

Target population

The current clinical practice guideline applies to all individuals under 18 years who may benefit from or are currently using HMV. Specific examples include individuals with spinal muscular atrophy (SMA), congenital and acquired central hypoventilation syndromes (CHS), chronic pulmonary disease such as bronchopulmonary dysplasia (BPD) and cystic fibrosis (CF), kyphoscoliosis, obesity hypoventilation syndrome (OHS), Duchenne muscular dystrophy (DMD), muscular dystrophies (MDs) other than DMD, myopathies and myotonic dystrophies.

Target users

Similar to the adult guidelines, this Clinical Practice Guideline on HMV in Children is intended to be a resource for:

a. physicians and healthcare providers caring for these children, not only within the tertiary care centres where ventilation is usually initiated, but also within the community, where these children will be primarily managed.

b. healthcare administrators (hospital and community) and related policy makers, to inform them of the issues involved, and what resources need to be in place for home ventilation to be a success, while providing maximum support and minimizing costs to the children and their families.
c. children and their families at risk for, or currently using, ventilatory support in the home, to be a resource and guide to inform them of the common problems met by these children, and suggested ways to address these problems.

Developed by The Canadian Thoracic Society (CTS), these guidelines are intended to provide the most up-to-date information and evidence-based recommendations to enable practitioners to manage the provision of HMV, both invasive and non-invasive, in children.

As noted in the adult guidelines, there are few prospective or randomized trials relevant to the management of adults on HMV; there are even fewer in children. Most of the recommendations are therefore based on retrospective or descriptive studies, frequently extrapolated from studies in adults, and, to a great extent, based on consensus among members of the CTS Paediatric HMV committee. The recommendations strive to achieve a balance between an optimal standard of care and the reality of healthcare in Canada, where geographic and economic barriers, as well as local availability of healthcare resources (such as polysomnography or trained home-care nursing/respiratory therapists) may limit what is achievable.

In addition, some issues considered important by the Committee are not addressed in the literature. For instance some jurisdictions have access to provincial ventilator pools where equipment and knowledgeable healthcare professionals are available at an affordable cost to ensure the success of HMV. Nonetheless, the literature does not address the presence or absence of government-funded equipment pools. Similarly, the "minimum" level of equipment required by individual patients (such as the necessity for back-up ventilators) remains a matter of opinion (and available resources), rather than being evidence-based.

Provision of HMV for children is a complex, interdisciplinary component of respiratory care and clinical practice. This component requires a continuum of chronic disease management involving many layers of expertise, from government and professional education to home-care services, acute and chronic health facilities and independent living facilities. The goal of HMV is to ensure the continued health of patients at risk for, and currently using, ventilatory support in their homes, where quality of life of the child and family is improved and the costs to the healthcare system are reduced.

**Methodology**

**Guideline development**

This clinical practice guideline was developed using a similar approach to the adult document, in accordance with the convention of the 23-item AGREE II instrument — the current gold standard in the appraisal of practice guidelines. The HMV Expert Committee comprised respirologists, a pediatrician, and a nurse practitioner from across Canada with content expertise in each of the topic areas. A methodologist conducted a systematic review of the literature that was current to June 2011. Before completion, the guideline was distributed to content experts in Canada and other countries with similar programs for the opportunity to provide feedback concerning the collection and interpretation of the evidence, as well as the development and content of the recommendations. Final consensus on the recommendations by the CTS HMV Committee was reached through a formal voting process that was anonymized. The literature will be reviewed biannually and the guideline will be updated as new or compelling evidence is identified.

**Literature search strategy**

The search strategy was designed to address questions related to broad issues with HMV, such as approaches to airway clearance and making the transition to home, and also to inform each sub-section of the HMV guideline addressing specific patient populations.

MEDLINE (OVID: 1980 through June 2011), Embase (OVID: 1980 through June 2011), HealthStar (1980 through June 2011) the Cochrane Library (OVID; Issue 1, 2009), the Canadian Medical Association InfoBase, and the National Guideline Clearinghouse were used to search the literature. Reference lists of related papers and recent review articles were also scanned for additional citations. A subsequent literature search was performed to update the literature review up until August 24, 2015, inclusive. Major relevant guidelines published during the review period in 2016 were also incorporated into the document.

The literature search of the electronic databases combined the following MeSH heading terms and text search terms to identify the body of published evidence on HMV related to the following disease related conditions: Lung Diseases/or Lung Diseases, Obstructive/or Chronic Obstructive Pulmonary Disease/or COPD.mp. or Neuromuscular Diseases/or Respiratory Insufficiency/or respiratory insufficiency.mp. or respiratory failure/or respiratory failure.mp. or breathing failure.mp. or breathing difficult*.mp. or respiratory muscle weakness.mp. or ("pulmonary function" and failure).mp. or Respiration Disorders/or Respiration/or Hypercapnia/or pulmonary disease.mp. or spinal muscle atrophy.mp. or Obesity Hypoventilation Syndrome/or OHS.mp. or Hypoventilation/or hypoventilation.mp. or muscular dystrophy.mp. or Muscular Dystrophies/or Kyphoscoliosis/or Kyphoscoliosis.mp. or neuromuscular disease/AND Treatment Related Terms: artificial ventilation/or ventilator/or ventilated patient/or Oxygen therapy/or assisted ventilation/or Ventilators, Mechanical/or Ventilation/or Ventilators, Negative Pressure/or ventilators negative pressure.mp. or ventil$.ti. or mechanical ventilation.mp. or Positive-Pressure Respiration, Intrinsic/or Intermittent Positive Pressure Ventilation/or Pulmonary Ventilation/or positive-pressure respiration.mp. or Positive-Pressure Respiration/or NIV.mp. or NIPPV.mp. orVA1.mp. or Respiration, Artificial/AND Health Care Setting Terms:Family centered care/or home care/or Home Care
Table 1. Grading recommendations.

<table>
<thead>
<tr>
<th>Grade of recommendation/Description</th>
<th>Benefit vs risk and burdens</th>
<th>Methodological quality of supporting evidence</th>
<th>Implications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A/ strong recommendation, high-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens, or vice versa</td>
<td>RCTs without important limitations or overwhelming evidence from observational studies</td>
<td>Strong recommendation, can apply to most patients in most circumstances without reservation</td>
</tr>
<tr>
<td>1B/ strong recommendation, moderate-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens, or vice versa</td>
<td>RCTs with important limitations (inconsistent results, methodological flaws, indirect or imprecise) or exceptionally strong evidence from observational studies</td>
<td>Strong recommendation, can apply to most patients in most circumstances without reservation</td>
</tr>
<tr>
<td>1C/ strong recommendation, low-quality or very-low-quality evidence</td>
<td>Benefits clearly outweigh risk and burdens, or vice versa</td>
<td>Observational studies or case series</td>
<td>Strong recommendation but may change when higher-quality evidence becomes available</td>
</tr>
<tr>
<td>2A/ weak recommendation, high-quality evidence</td>
<td>Benefits closely balanced with risks and burden</td>
<td>RCTs without important limitations or overwhelming evidence from observational studies</td>
<td>Weak recommendation, best action may differ depending on circumstances or patients’ or social values</td>
</tr>
<tr>
<td>2B/ weak recommendation, moderate-quality evidence</td>
<td>Benefits closely balanced with risks and burden</td>
<td>RCTs with important limitations (inconsistent results, methodological flaws, indirect or imprecise) or exceptionally strong evidence from observational studies</td>
<td>Weak recommendation, best action may differ depending on circumstances or patients’ or social values</td>
</tr>
<tr>
<td>2C/ weak recommendation, low-quality or very-low-quality evidence</td>
<td>Uncertainty in the estimates of benefits, risks and burden; benefits, risks and burden may be closely balanced</td>
<td>Observational studies or case series</td>
<td>Very weak recommendations; other alternatives may be equally reasonable</td>
</tr>
</tbody>
</table>

Services/or home.ti. or home ventilat$.mp. or (home.mp. and Long-Term Care/) or Home care.mp. orhomecare.mp. 6 orHMV.mp. or home ventil$.mp. or (assisted living and ventilator).mp. or ventilator assisted living.mp. or (home ventil* and Mechanical).mp. or (home ventil* and machi-ne*).mp. or (oxygen equipment and home).mp. or home ventilation program*.mp. or home mechanical ventilation.ti. AND Limits: Humans, English, All Children: 18 years and younger.

Study selection criteria

Articles were selected for inclusion in the systematic review of the evidence if they reported data on the role of HMV among children who require ventilatory assistance. More specifically, articles that met this criterion must have addressed the question: Does HMV lead to better patient, caregiver or system outcomes than other currently available ventilation and management options (e.g., ventilation in a chronic care facility, such as a hospital, or no mechanical ventilation)? Studies were required to report data on at least one of the following outcomes of interest: survival, pulmonary function, sleep parameters, airway clearance techniques, cognition, VAI’s and caregiver’s QoL, monitoring at home, making the transition to home, making the transition to adult care, or ethical considerations.

In the event of modest data, it was agreed that expert consensus would be used to form the recommendations. As such, only the highest levels of evidence considered sufficient to inform recommendations were chosen for each subsection of the guideline. In descending order of preference, minimum levels of evidence were gathered to inform the clinical questions. Sources included were clinical practice guidelines, systematic reviews, meta-analyses, randomized controlled trials, non-randomized comparative studies, prospective single-cohort studies, retrospective single-cohort studies and case studies. Given the limited utility of the systematic review, guideline authors were at liberty to perform their own systematic review (up until December 31, 2013), to identify any literature relevant to their section. Articles were excluded from the systematic review of the evidence if they were reported in a language other than English.

Critical appraisal

The strengths and weaknesses of the evidence were carefully considered in the generation of the recommendations. Although the majority of the evidence on this topic area is modest, the Grading of Recommendations Assessment, Development and Evaluation (GRADE) methodology was used to inform the generation of recommendations and critically appraise the strength of the evidence. Where no evidence was available, the committee made a consensus-based recommendation and the recommendation was identified as such (Table 1).

Organization of results

This guideline is divided into separate sub-sections based either on specific patient populations (six sub-sections) or specific issues common to all patients on HMV (six sub-sections). Each section describes the literature findings provides key recommendations and the supporting level of evidence and where possible, identifies barriers to implementation of recommendations.
Disclosure statement
No potential conflict of interest was reported by the author(s).

References
