

## An Official American Thoracic Society Clinical Practice Guideline: Pediatric Chronic Home Invasive Ventilation

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**Background:** Children with chronic invasive ventilator dependence living at home are a diverse group of children with special health care needs. Medical oversight, equipment management, and community resources vary widely. There are no clinical practice guidelines available to health care professionals for the safe hospital discharge and home management of these complex children.

**Purpose:** To develop evidence-based clinical practice guidelines for the hospital discharge and home/community management of children requiring chronic invasive ventilation.

**Methods:** The Pediatric Assembly of the American Thoracic Society assembled an interdisciplinary workgroup with expertise in the care of children requiring chronic invasive ventilation. The experts developed four questions of clinical importance and used an evidence-based strategy to identify relevant medical evidence. Grading of Recommendations Assessment, Development, and Evaluation (GRADE) methodology was used to formulate and grade recommendations.

**Results:** Clinical practice recommendations for the management of children with chronic ventilator dependence at home are provided, and the evidence supporting each recommendation is discussed.

**Conclusions:** Collaborative generalist and subspecialist comanagement is the Medical Home model most likely to be successful for the care of children requiring chronic invasive ventilation. Standardized hospital discharge criteria are suggested. An awake, trained caregiver should be present at all times, and at least two family caregivers should be trained specifically for the child's care. Standardized equipment for monitoring, emergency preparedness, and airway clearance are outlined. The recommendations presented are based on the current evidence and expert opinion and will require an update as new evidence and/or technologies become available.

**Keywords:** respiratory insufficiency; mechanical ventilators; discharge planning; medical home; children

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## Overview

The goal of this guideline is to provide an approach, based on the available evidence and the consensus of the assembled group of experts, to the hospital discharge and subsequent management of children using invasive ventilation in the home. The guideline is intended for use by pediatric pulmonologists, pediatric intensive care physicians, pediatricians, family physicians, pediatric nurse practitioners, pediatric physician assistants, pediatric nurses (both in the hospital and in the home), and respiratory therapists who care for these patients. The recommendations made in the guideline are not intended to impose a standard of care. Children requiring invasive ventilation are a diverse group of patients with complex health care needs. There are a wide variety of unique and important social and medical issues that will need to be considered when planning for the care of each individual patient. It is our hope that the guideline presented can provide practitioners a basis for sound decision making and safe, effective care for this group of children.

An interdisciplinary Pediatric Chronic Home Ventilation Workgroup was convened by the Pediatric Assembly of the American Thoracic Society to develop a guideline to facilitate comprehensive home management of respiratory technology-dependent children and highlight areas for future research. Four key clinical questions were identified by the Workgroup and addressed in the guideline. The recommendations formulated are based on the indirect evidence in the available literature, which consisted of uncontrolled studies or observations and the consensus of the Workgroup members. Grading of Recommendations Assessment, Development and Evaluation (GRADE) methodology was used to assess the quality of the published evidence and evaluate the strength of the recommendations. The following recommendations were made by the Workgroup:

1. For children requiring chronic home invasive ventilation, we suggest a comprehensive Medical Home comanaged by a generalist and respiratory subspecialist (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
2. For children requiring chronic invasive ventilation, we suggest the use of

standardized discharge criteria to objectively assess readiness for care in the home (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).

3. We recommend that an awake and attentive trained caregiver be in the home of a child requiring chronic invasive ventilation at all times (Strength of Recommendation: Strong; Quality of Evidence: Very Low).
4. For children requiring chronic invasive ventilation, we suggest that at least two specifically trained family caregivers are prepared to care for the child in the home (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
5. We suggest that ongoing education to acquire, reinforce, and augment skills required for patient care be provided to both the family and professional caregivers of children requiring chronic home invasive ventilation (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
6. For children requiring chronic home invasive ventilation, we suggest monitoring, especially when the child is asleep or unobserved, with a pulse oximeter rather than use of a cardiorespiratory monitor or sole use of the ventilator alarms (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
7. For children requiring chronic home invasive ventilation, we recommend regular maintenance of home ventilators and all associated equipment as outlined by the manufacturer.
8. We suggest the following pieces of equipment for use in the home when caring for a patient on home mechanical ventilation: the ventilator, a back-up ventilator, batteries, a self-inflating bag and mask, suctioning equipment (portable), heated humidifier, supplemental oxygen for emergency use, nebulizer, and a pulse oximeter (nonrecording) (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
9. We suggest that a mechanical insufflation-exsufflation device be used to help maintain airway patency in patients requiring home mechanical ventilation with ineffective cough, including, but not limited to, those with neuromuscular disease with poor

respiratory muscle strength (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).

## Introduction

Of the more than 9.4 million children in the United States with special health care needs, few present with more challenges than those faced by children requiring chronic home invasive ventilation (1–3). The shift from care in the pediatric intensive care unit within the hospital to intensive care at home requires invested families, multiple trained in-home caregivers, sophisticated technology, financial support, and ready access to primary and subspecialty medical care. Despite the mandate for national action by the Institute of Medicine in 2003 for children with special health care needs (CSHCN), there are no evidence-based guidelines for this complex subgroup of technology-dependent children (4). The Pediatric Assembly of the American Thoracic Society (ATS) proposed the development of an evidence-based clinical practice guideline for management of children requiring invasive mechanical ventilation at home. An interdisciplinary Pediatric Chronic Home Ventilation Workgroup was convened to develop a guideline to facilitate comprehensive home management of respiratory technology-dependent children and highlight areas for future research. The participants in this guideline include experts in pediatric sleep medicine and pulmonary and critical care, ATS assembly chairs, and physician and nursing experts in the inpatient and outpatient care of children with advanced respiratory needs.

We recognize that children requiring invasive ventilation are a diverse group of patients. These patients have advanced pulmonary disease, neuromotor disease, or defects in respiratory drive with the common consequence being insufficient minute ventilation to maintain normocapnia and/or adequate oxygen saturations. They also range in severity, from children who have inadequate respiration solely during sleep, hence requiring nocturnal respiratory assistance alone, to those who require ventilation around the clock for survival. Despite this heterogeneity, common themes and challenges are found. The Workgroup investigated four areas that are critical in

the care of these patients, addressing coordination of care, readiness for home care, training of caregivers, and necessary equipment, and made recommendations on the basis of available literature and consensus of the Workgroup experts.

The goal of this guideline is to provide an approach, on the basis of the available evidence and the consensus of the assembled group of experts, to the hospital discharge and subsequent management of children using invasive ventilation in the home. The guideline is intended for use by pediatric pulmonologists, pediatric intensive care physicians, pediatricians, family physicians, pediatric nurse practitioners, pediatric physician assistants, pediatric nurses (both in the hospital and in the home), and respiratory therapists who care for these patients. The recommendations made in the guideline are not intended to impose a standard of care. There are a wide variety of unique and important social and medical issues that will need to be considered when planning for the care of each individual patient. It is our hope that the guideline presented herein can provide practitioners a basis for sound decision making and safe, effective care for this group of children.

## Questions

The key clinical questions identified by the Workgroup as important for target users are:

1. Should the Medical Home or family-centered care (FCC) concepts be used in the care of children requiring chronic home invasive ventilation?
2. Should standardized discharge criteria be used when planning the hospital discharge of children requiring chronic home invasive ventilation?
3. Should home caregivers be specifically trained in the care of children requiring chronic home invasive ventilation?
4. Should standard home equipment requirements be applied when planning for the equipment needs of children requiring invasive ventilator support in the home?

## Methods

These clinical practice guidelines were prepared according to the methods of the ATS, which are described in detail in the online supplement and summarized in

Table 1. The Patient/Population, Intervention, Comparison, and Outcome (PICO) framework was used to search systematically for relevant evidence. The searches identified no published randomized controlled trials or controlled observational studies. The recommendations are, therefore, based on uncontrolled studies and the clinical observations of the Workgroup members. Grading of Recommendations Assessment, Development, and Evaluation (GRADE) methodology was used to assess the quality of the published evidence and rate the strength of the recommendations (5). All evidence that was selected to inform the Workgroup's decisions was characterized as very low quality, indicating that the estimated effects of our suggested interventions are uncertain and very likely to be affected by future research. The strength of one recommendation was rated as strong, and all others were rated as conditional; the appropriate interpretation of the strength of a recommendation is provided in Table 2 (5). The recommendations are detailed in Table 3.

## Clinical Questions and Recommendations

### Question 1: Should the Medical Home or Family-centered Care Concepts Be Used in the Care of Children Requiring Chronic Home Invasive Ventilation?

**Background.** The Workgroup believed that incorporating the concepts of FCC and the Medical Home into the care of the medically complex children requiring chronic home invasive ventilation is a priority. The Workgroup found no published experimental or observational studies examining the effect of using FCC and Medical Home models on outcomes in children requiring invasive ventilation in the home. There is literature describing and examining these models in children with complex health care needs, and this is reviewed in the following section. On the basis of the clinical experience of the Workgroup and the available indirect literature reviewed, we suggest using a comprehensive Medical Home comanaged by the generalist and respiratory subspecialist for children requiring chronic home invasive ventilation. This recommendation places

high value on the possible medical and social benefits and low value on the potential risks, which may include increased resource use and provider time. Structure and components of the comanaged Medical Home are described later and in Table 4.

**CSHCN.** In 1998, CSHCN were defined by the Maternal and Child Health Bureau (MCHB) of the U.S. Health Resources and Services Administration as follows:

CSHCN are those who have or are at increased risk for a chronic physical, developmental, behavioral, or emotional condition and who also require health and related services of a type or amount beyond that required by children generally (6).

The "health and related services" referenced by the above definition include physician subspecialists, hospitals specialized in the care of children, therapeutic services (e.g., speech, physical, occupational, and others), home health services, home nursing services, family support services such as family counseling, comprehensive case management, care coordination, specialized equipment and supplies, and related services such as social services, transportation, special education, medical daycare, early intervention, and others. Children with a tracheostomy on long-term ventilatory support clearly qualify as CSHCN and, indeed, represent the extreme end of the spectrum with respect to their complexity, intensity of care requirements, service needs, and risk of adverse outcome in their daily lives.

The definition of CSHCN does not require a specific diagnosis or condition; it is intentionally broad and inclusive of a wide spectrum of medical conditions. The intention was to assist and guide public programs and other organizations in program planning. From its inception, this definition has been used in program development by MCHB, U.S. Health Resources and Services Administration, and state Title V programs for needs assessment and to facilitate meeting federal requirements when developing a community system of services for CSHCN (6).

**Location of care: home versus institutional care.** Home care for the majority of ventilator-dependent children is a relatively recent development. In the United States, long-term care of ventilator-dependent children with their family, in the home environment, dates from the 1970s

**Table 1.** Summary of Methodology

Method	Yes	No
Panel assembly		
Included experts for relevant clinical disciplines	X	
Included individuals who represent the views of patients and society at large	X	
Included a methodologist with appropriate expertise (documented expertise in conducting systematic reviews to identify the evidence base and the development of evidence-based recommendations)	X	
Literature review		
Performed in collaboration with librarian	X	
Searched multiple electronic databases	X	
Reviewed reference lists of retrieved articles	X	
Evidence synthesis		
Applied prespecified inclusion and exclusion criteria	X	
Evaluated included studies for sources of bias	X	
Explicitly summarized benefits and harms		X
Used PRISMA1 to report systematic review		X
Used GRADE to describe quality of evidence	X	
Generation of recommendations		
Used GRADE to rate the strength of recommendations	X	

Definition of abbreviations: GRADE = Grading of Recommendations Assessment, Development, and Evaluation; PRISMA1 = Preferred Reporting Items for Systematic Reviews and Meta-Analysis 1.

and 1980s, when portable ventilators became practical for home use. A Surgeon General’s Department of Health and Human Services workshop on ventilator-dependent children, held in 1982, enumerated principles that included “the full participation of the parents and siblings in all the processes of medical care” and “linking up the child and the family with available social and medical support groups in the community,” with a major goal of developing “strategies for providing community-based services as an alternative to acute care facilities” (2). A major shift away from institutional care was given national recognition by the U.S. Surgeon General’s 1987 commitment to “family-centered, community-based, coordinated

care” for CSHCN (3). The Surgeon General’s report indicated that CSHCN “deserve to live with their families in their own communities, and to share in the everyday experiences most Americans take for granted. We must make a commitment to provide the kinds of health care these children require, in ways that allow them to participate as fully as possible in all aspects of family and community life.” The United States national objectives for improving health, Healthy People 2010 and 2020, both have explicit goals of reducing the number of children and youth with disabilities living in congregate care residences (7).

Hospital pediatric intensive care units, acute-care wards, and even chronic

residential care facilities are not designed for family-centered child development, schooling, or family and social life or to meet the long-term developmental needs of a child. Thus, for decades U.S. government health agencies have accepted as core principles that the best place for the wellbeing of a technology-dependent child is with the family/caregivers in the community, and that all aspects of the medical care should be family centered, coordinated, and integrated with community services (2, 3).

**FCC.** At its core, FCC of a child is a partnership approach to decision making between health professionals and the family in all aspects of care (8). The basic principles of FCC shared by all current definitions include 1) unbiased, open information sharing with the patient and family; 2) a working relationship with the family that honors and respects differences; 3) collaborative decision making that includes families as partners; 4) negotiation of medical care plans and desired outcomes with the family; and 5) all decision making and medical care within the context of the child’s family, home, daily activities, school, and life in the community (8). FCC and the closely related patient-centered care approach have been endorsed by multiple U.S. government health care agencies including MCHB, multiple medical societies, Healthy People 2020, and the Institute of Medicine. In a 2012 Policy Statement, the American Academy of Pediatrics (AAP) recommended incorporation of the core concepts of patient- and family-centered care into all aspects of pediatric medical care and defines the patient and family as integral members of the health care team (9). Numerous benefits of FCC have been demonstrated for patients and families

**Table 2.** Interpretation of Strong versus Conditional Strength of Recommendation

User Group	Strong Recommendation	Conditional Recommendation
Patients	Most individuals in this situation would want the recommended course of action and only a few would not. Formal decision aids are not likely to be needed to help individuals make decisions consistent with their values and preferences.	The majority of individuals in this situation would want the suggested course of action, but many would not.
Clinicians	Most individuals should receive the intervention. Adherence to this recommendation according to the guideline could be used as a quality criterion or performance indicator.	Understand that different choices will be appropriate for individual patients. Decision aids may be useful in helping individuals make decisions consistent with their values and preferences.
Policy makers	The recommendation can be adopted as policy in most situations.	Policy making will require substantial debates and involvement of many stakeholders.

**Table 3.** Summary of Recommendations

Recommendation	Strength	Quality of Evidence	Remarks
1. For children requiring chronic home invasive ventilation, we suggest a comprehensive Medical Home comanaged by the generalist and respiratory subspecialist.	Conditional	Very low	The Medical Home model can provide family-centered care for children with special health care needs, including children on home invasive ventilation. This recommendation places a high value on the possible medical and social benefits to this intervention and places low value on the potential risks, which may include increased provider time.
2. For children requiring chronic invasive ventilation, we suggest the use of standardized discharge criteria to objectively assess readiness for care in the home.	Conditional	Very low	The Workgroup believed that comprehensive standardized discharge criteria would encourage a complete review of each patient’s medical stability and home situation to facilitate safe discharge. The goal is to identify and eliminate important barriers to care in the home before discharge and consider alternate care arrangements if obstacles cannot be eliminated. Weight given to each component of the proposed criteria would vary from patient to patient. The recommendation places high value on the potential benefits of considering all facets of a child’s care in the home before discharge and low value on the increased provider time and resource use that may be required.
3a. We recommend that an awake and attentive trained caregiver be in the home of a child requiring chronic invasive ventilation at all times.	Strong	Very low	Despite very low quality evidence supporting this recommendation, the Workgroup was confident that in this case the desirable consequences would clearly outweigh the undesirable consequences of following this recommendation. Lack of an awake and attentive trained caregiver would place the child in a life-threatening situation. Training of caregivers is irrelevant if one is not available to respond to an emergent situation. For most families this requires the support of a professional appropriately trained in-home caregiver to allow family caregivers time to sleep, work, and maintain a life balance. This recommendation places a high value on the safety of the patient, and low value is placed on avoiding the increased use of resources and the possible disruption to families who may need to accommodate a professional caregiver in their home.
3b. For children requiring chronic invasive ventilation, we suggest that at least two specifically trained family caregivers are prepared to care for the child in the home.	Conditional	Very low	The experience of the Workgroup and available data indicate that a lone trained family caregiver would rarely be capable of shouldering the entire burden of care for a child using invasive ventilation in the home. This recommendation places high value on the safety of the patient and quality of life of caregivers and low value on increased resource use for training more than one caregiver.
3c. We suggest that ongoing education to acquire, reinforce, and augment skills required for patient care be provided to both the family and professional caregivers of children requiring chronic home invasive ventilation.	Conditional	Very low	The Workgroup believed, based on clinical experience, that practitioners and professional personnel agencies must strive to provide ongoing education to family and professional caregivers. Continuing education would help reinforce learned skills and allow training on new technologies and protocols. This recommendation places a high value on safety and on the potential clinical benefits to the patient and a low value on increased cost and resource use.
4a. For children requiring chronic home invasive ventilation, we suggest monitoring, especially when the child is asleep or unobserved, with a pulse oximeter rather than use of a cardiorespiratory monitor or sole use of the ventilator alarms.	Conditional	Very low	Small indirect studies and the experience of the Workgroup suggest that ventilator alarms may not always function correctly. Furthermore, hypoxemia is most likely to be the first indicator of a serious issue in a child with respiratory disease. The workgroup believes pulse oximetry is the preferred method for monitoring patients on home mechanical ventilation. This recommendation places high value on the safety of the child and low value on possible increase in caregiver burden secondary to false alarms.

(Continued)

Table 3. (Continued)

Recommendation	Strength	Quality of Evidence	Remarks
4b. For children requiring chronic home invasive ventilation, we recommend regular maintenance of home ventilators and all associated equipment as outlined by the manufacturer.			Although states have differing regulatory requirements for DME providers, and the data supporting the value of equipment maintenance are lacking, the Workgroup believed strongly that maintenance of all home equipment by appropriately trained DME employees as recommended by the manufacturer should be standard of care. Care should be taken to assure that the actual ventilator settings as seen on the control panel match the prescribed settings. Twenty-four hour a day service and phone support must be available. This recommendation places a high value on the likely clinical benefits of properly functioning equipment programmed with the correct patient settings and low value on increased resource use.
4c. We suggest the following pieces of equipment for use in the home when caring for a patient on home mechanical ventilation: the ventilator, a back-up ventilator, batteries, a self-inflating bag and mask, suctioning equipment (portable), heated humidifier, supplemental oxygen for emergency use, nebulizer, and a pulse oximeter (nonrecording).	Conditional	Very low	On the basis of experience, the Workgroup believed the presence of specific pieces of equipment could prevent the development of life-threatening situations and/or reduce their severity. This recommendation places high value on the potential to avoid emergent situations due to the presence of important reserve and emergency equipment and low value on increased resource use and increased equipment costs.
4d. We suggest that a mechanical insufflation–exsufflation device be used to help maintain airway patency in patients requiring home mechanical ventilation with ineffective cough, including, but not limited to, those with neuromuscular disease with poor respiratory muscle strength.	Conditional	Very low	Equipment to facilitate airway clearance is essential in reducing the risk of acute airway obstruction in patients with ineffective cough. This recommendation places high value on the potential to avoid emergent airway plugging and low value on increased costs and resource use.

Definition of abbreviation: DME = durable medical equipment.

and for physicians who practice patient- or family-centered care, including CSHCN (9–11). There are no studies to date of patient- or family-centered care specifically for children receiving long-term mechanical ventilation.

**The Medical Home.** The Medical Home is the best-known model of health care delivery for CSHCN. It is important to note that it is not a specific geographical location, medical office, or provider. The AAP describes the Medical Home concept as care that is 1) accessible, 2) comprehensive, 3) compassionate, 4) continuous, 5) coordinated, 6) culturally competent, and 7) family centered (12). Most concepts of the Medical Home feature a primary Medical Home provider, and it is often assumed that it must be a primary care physician. However, there are no requirements that the primary Medical Home provider be a primary care physician. Similarly, there are no requirements concerning where it is based; as long as the Medical Home meets the criteria above, it can be primary care based, tertiary care based, or a primary–tertiary

comanagement hybrid model. However, the AAP is clear and specific on the point that Medical Home care “should be delivered or directed by well-trained physicians who provide primary care and help to manage and facilitate essentially all aspects of pediatric care” (13). Numerous state and federal laws mandate community-based, family-centered, coordinated care programs for CSHCN, and the AAP and MCHB strongly endorse the Medical Home comprehensive care model as the preferred approach for all children, including CSHCN (12, 14, 15).

**Care models for children receiving chronic invasive mechanical ventilation.** As outlined above, in the United States, FCC is already mandated for CSHCN, and the Medical Home concept is the recommended model of comprehensive care coordination for CSHCN. Given that children dependent on invasive, long-term mechanical ventilation clearly are CSHCN, these approaches to health care delivery and coordination are already mandatory for this population. Thus, the question for this group of children, at the extreme end of the

complexity spectrum and dependent on highly specialized, complicated life-sustaining technology, remains: What is the best model for delivering Medical Home care? Should primary Medical Home management for the ventilator-dependent child be provided by the primary care physician, the tertiary care pulmonologist, a hybrid model in which both the primary care and subspecialty physician act as Co-Medical Home providers, or a hospital-based comprehensive Medical Home clinic? The remainder of this section discusses the evidence for a Medical Home concept and considers possible models of Medical Home care delivery.

**Care needs of the ventilator-dependent child: a complex mix of specialty and primary care.** The care, equipment, supply, and monitoring needs specific to the ventilator-dependent child are outlined in the following sections of this document. The required technology always includes the tracheostomy, ventilator, humidifier, and suction devices and often includes added technology such as airway clearance devices, mechanical insufflation–exsufflation (MI-E)

**Table 4.** Features of a Comanaged Medical Home for Children Requiring Chronic Invasive Ventilation in the Home**The care would be family/patient centered.**

- Recognition of patient/family preferences, social services availability, barriers to communication or medical provision is necessary.

**Medical Home provider oversight would be provided by a collaborative partnership between the generalist and pediatric pulmonologist, and other necessary subspecialists.**

- Keeping in mind that many children requiring mechanical ventilation are equally dependent on subspecialists, such as neurologists, gastroenterologists, physical therapists, etc., this collaborative partnership depends on the situation and will require oversight from a specific provider most accessible to the family and identified as primarily accountable. This provider is frequently but not necessarily the primary care provider.

**The specific roles and responsibilities of each provider would be clearly delineated for each practitioner, all members of both teams, and the patient and family.**

- In practice, a written summary of responsibilities will be provided to the team and family by a social worker or nurse or primary care provider outlining expectations and order of communication channels.

**The pulmonologist and team would be responsible for management of all pulmonary and related aspects of care.****The generalist would be responsible for all aspects of primary care.****The comanagement collaborative would decide and delineate responsibility for comprehensive assessment, coordination, and management of all other aspects of care.**

- This includes access to medical care, transportation, family care and respite, access to nutritional needs, community resources, etc. A social worker or local public health resource may be crucial to assist the primary care provider. Pulmonologists, medical specialists, and their teams will be required to assist in coordination of complex care requirements and appointments to lessen the burden on the family. Much care can be delegated to the local primary caregivers with communication from the specialist teams.

**Effective communication tools would be used to ensure effective implementation of the comprehensive care plan, without redundancy or duplication.**

- Ideally, access to the same electronic medical record system for communication between the primary care provider, specialists, nursing staff, and social workers would allow sharing of family communications, expected appointments, and ongoing medical issues. If this electronic medical record system is not available to all providers, communications should be outlined in writing to the comanagement collaborative.

devices, supplemental oxygen, monitoring devices such as pulse oximeters, and, typically, numerous medications that require the use of aerosol delivery devices. As outlined in sections below, continuous 24 h/d, 7 d/wk monitoring is required for these children due to the high risk of morbidity and mortality (16, 17). Therefore, skilled care by trained caregivers, usually a combination of family members with home nursing services when available, is required.

Children dependent on long-term mechanical ventilation are at risk for several complications directly related to their chronic pulmonary condition, including pulmonary hypertension, recurrent pulmonary infections, poor growth, developmental delay, and life-threatening emergency events such as tracheostomy plugging, accidental decannulation, and other causes of cardiorespiratory arrest. Thus, caregivers (the parents and homecare providers) must be able to perform all routine care, recognize and be able to troubleshoot problems, and have the skill and training to respond effectively. Oversight of the care outlined above is typically considered the primary responsibility of the pulmonologist and pulmonary care team. The degree of involvement of otolaryngology for tracheostomy care and airway issues or of

cardiology for issues related to pulmonary hypertension varies between states and institutions.

Many ventilator-dependent children also have other major medical conditions such as neurological disorders (e.g., cerebral palsy, central nervous system injury, congenital central hypoventilation), neuromuscular disorders (e.g., muscular dystrophy), congenital anomalies (including a wide variety of genetic syndromes), and/or congenital heart disease (17, 18). These comorbid conditions each may have its own set of intensive health care needs and may involve dependence on nonrespiratory technologies (e.g., renal dialysis, cardiac pacemaker, ventriculoperitoneal shunt, baclofen pump) involving different subspecialists, such as gastroenterology, cardiology, otolaryngology, neurology, and others. Frequently, the ventilator-dependent child also has chronic dysphagia, with limited oral intake and dependence on liquid feedings via gastrostomy or gastrojejunal feeding tube. Some of the comorbid conditions may interact; a common example is dysphagia leading to recurrent/chronic aspiration and resulting in adverse effects on respiratory health.

Children on long-term ventilation should also receive speech therapy, physical

therapy, and occupational therapy. They may require evaluation and therapies for developmental delay and behavioral issues. In the educational sphere, the family of the ventilator-dependent child interacts at multiple levels with schools, including school administrators, nurses, teachers, special education teachers, and sometimes therapists. School attendance outside the home for the child on long-term invasive ventilation is complicated. In addition to the ventilator, suction devices, other tracheostomy care supplies, and monitoring equipment, the child may be on supplemental oxygen. Children with tracheostomy must have a trained caregiver present at all times, even in school. Many, if not most, require specialized wheelchairs and other adaptive equipment and have complex transportation requirements. Families must deal continuously with the legal and financial aspects of this complex, high-intensity, high-cost home care.

In addition to all of the above services, the ventilator-dependent child requires primary care services common to all children, including immunizations, well-child care, anticipatory guidance, and evaluation and treatment for common childhood infections and other disorders.

**Medical care for the ventilator-dependent child: family stress, frustration, and disruption.** For CSHCN, care coordination has been defined as “a process that links children with special health care needs and their families to services and resources in a coordinated effort to maximize the potential of children and provide them with optimal health care” (19). At the present time, health care for the ventilator-dependent child is a complex mix of primary care and subspecialty care needs involving multiple disciplines, with care coordination often defaulting to the family. Families often cite care coordination and navigating the complex, fragmented health care system as major stressors in their daily lives (20).

In the care of children with special health care needs, the specific roles and responsibilities of the primary care and subspecialty physicians and their teams are often not clearly delineated. Care coordination and communication can be complex, confusing, and frustrating for families, especially in children requiring care from multiple subspecialists. Where is the Medical Home based? Which health care practitioner is the primary Medical Home provider? Who takes responsibility for comprehensive care coordination? What are the specific responsibilities of each subspecialist involved in the child’s care? Who monitors communication between all involved services?

Medical complexity is a major factor determining the quality of care in CSHCN. Indeed, of CSHCN, medically complex children (those requiring special medical equipment such as tracheostomy or gastrostomy tubes) have higher unmet needs in health care and community-based services, such as assistive devices and special therapies (21). Focus group discussions conducted with regional coordinators of early intervention programs in Massachusetts identified significant common issues in medically complex children requiring mechanical ventilation (22). The lack of expertise of community caregivers leaves the training of staff to the parents, who justifiably worry about child safety. Anxiety can increase over time with worry about sudden health changes (23). Families of children with respiratory insufficiency can become physically and socially isolated due to the difficulty finding family supports or sufficiently trained home-care workers

to provide respite care or time for work (24, 25).

Due to a nationwide shortage of nursing, unfilled nursing shifts at home become the parent’s responsibility (26, 27). Family stress, marital problems, and sleep deprivation are regular occurrences when caregiving is left to an unsupported family (28–31). Travel logistics and trouble with coordination of medical care is increased in families without an established Medical Home (24). With all of the constraints involved in caring for the medically complex child, there is a high rate of missed work, financial stress, and parental unemployment (32). Parents of CSHCN have higher rates of poor physical and emotional health and depression (33, 34). In families with a child affected by cerebral palsy, parents report higher systemic symptoms of illness and chronic conditions (35). Family-centered care provided via a Medical Home approach aims to reduce stress by providing care coordination, resources to help navigate the health care system, and community support (36).

**Challenges of care delivery for ventilator-dependent children.** As noted above, ventilator-dependent children represent an extreme on the severe end of the spectrum of children with medical complexity. A study of inpatient health care use data highlights some of the differences between ventilator-dependent children and other children with complex chronic health conditions. Children dependent on long-term ventilation had longer lengths of stay in the hospital, greater total costs, higher risk of death, greater use of home health care, and a greater likelihood of transfer to a nonacute facility (37). Although the number of children dependent on long-term ventilation has increased in recent years, there is a dearth of data regarding the epidemiology of these children and how they live and receive medical care outside of the acute care setting (16, 22, 37, 38).

The 2012 AAP clinical report entitled “Home Care of Children and Youth with Complex Care Needs and Technology Dependencies” delineates desirable features of any Medical Home for children with complex medical and developmental issues (18). Unfortunately, the reality of delivering or receiving that medical care often departs sharply from the coordinated and family-centered care recommended. Fragmented, uncoordinated, and inefficient care systems have been described frequently in the

medical literature (20, 23, 39). Pilot programs that provide Medical Homes and coordinate care have demonstrated improved outcomes and decreased overall health care expenditures (39–42). However, these programs operate at financial deficits and include children with complex chronic conditions in general, not specifically those who require long-term ventilation.

Information regarding the current patterns of health care delivery to ventilator-dependent children comes largely from informal discussion among experts and limited studies (22). In many cases, a pediatric pulmonary group serves some of the Medical Home functions, providing medical coverage and care coordination in the ambulatory, emergency department, inpatient, and home environment, 24 hours per day and 7 days per week. This role is similar to that of the special needs physician in the primary care–tertiary care partnership model described by Gordon and colleagues (39). A number of centers have chronic ventilation programs with dedicated nurse coordinators, usually housed in the pediatric pulmonary divisions. In major tertiary care centers, ambulatory visits to a pulmonologist often include additional visits with other members of the health care team, such as respiratory therapists, social workers, and nutritionists. In general, physicians spend 1 hour on each ambulatory visit but are often allotted much less time in the schedule. Physicians and their staff devote hours to each patient between visits.

Since the Institute for Healthcare Improvement’s landmark report on medical error in 1999, institutions have focused on improving systems and processes within hospitals to improve patient safety and quality of care (43). Particular emphasis has been placed on the critically ill patients within hospital intensive care units. Children requiring long-term mechanical ventilation are chronically critically ill (44). Delivering their medical care outside the acute care hospital requires a web of interactions among patients and families, multiple layers of physicians and allied health care professionals, and home care companies. When surveyed, the Workgroup members reported major problems with communication on medically critical issues such as: ventilator settings, home care orders, identification of which physician is responsible for which order, and medical records. Although

adverse medical and/or safety-related events within health care delivery systems are often tracked, there is no system to track or report such incidents in ventilator-dependent children once they leave the hospital.

**Care model for family-centered Medical Home care of ventilator-dependent children.** The complexity of the care required for a child dependent on chronic mechanical ventilation cannot be overstated. Regardless of the venue of care, a myriad of providers contribute to the development and implementation of the care plan. To achieve optimum functioning, this care plan must be comprehensive and address the child's medical, educational, psychosocial, and, often, behavioral needs. Care of this complexity is potentially high cost and draws on many community resources. Each specific area typically involves multiple providers or disciplines, and there is often overlap in responsibilities. What is the best, most practical and feasible way to provide family-centered, Medical Home care for the ventilator-dependent child?

Several models of shared care have been described. The first model, endorsed by the AAP for routine care, is the Medical Home with generalist as primary provider and input from specialists only occasionally as needed. Another model proposes comanagement by the generalist and subspecialist, both as Medical Home providers, each with clearly delineated roles. In a third model, the subspecialist is the primary Medical Home provider, with the generalist playing a limited role as needed (45). In a fourth proposed model, the hospital-based comprehensive care Medical Home clinic, all primary and specialty care and care coordination is provided in the tertiary care setting by a multidisciplinary team (40). As outlined by Antonelli and colleagues, the choice of model depends on 1) the medical complexity and severity of the medical condition(s), 2) the expertise of each type of provider, 3) family/patient access to each type of provider, and 4) the comfort level of families and involved providers in different care scenarios (45).

Regardless of the model, the Medical Home for the ventilator-dependent child must provide:

- Subspecialty management of the child's chronic respiratory failure and its treatment, including all aspects of

ventilator management and associated therapies

- Support for technical aspects of the ventilator and tracheostomy
- Appropriate training of family caregivers
- Access to care 24 h/d, 7 d/wk
- Community-based primary care
- Coordination of all aspects of the child's care
- Guidance to patient/family on all aspects of the child's medical care
- Providers responsible for comprehensive care assessment and management of all aspects of the child's health care

Given the extreme level of medical complexity of the child on long-term mechanical ventilation, with highly specialized care and medical technology, the generalist as primary Medical Home provider model is unlikely to be successful. This model would require the generalist provide and be responsible for a comprehensive care plan and management, with limited input from the subspecialist (45). Similarly, in view of the extensive nonpulmonary care coordination needs outlined above, the subspecialist as primary Medical Home provider model may also prove difficult to implement. This model would require the subspecialist to provide comprehensive care assessment and management of all aspects of the child's care, including nonpulmonary problems. It is unlikely that a pediatric pulmonologist would be comfortable, have the expertise, and have time or resources to oversee the primary care and other, nonpulmonary aspects of the child's care. The hospital-based comprehensive care Medical Home clinic model has been demonstrated to be successful (40), but this model of care exists only in a few locations; it assumes that pediatric pulmonologists, respiratory therapists, and related services are part of the multidisciplinary team caring for the patient; and it assumes that the family has easy access to the tertiary care medical center.

The model most likely to be successful for providing comprehensive Medical Home care to the ventilator-dependent child is the collaborative generalist and subspecialist comanagement model (39, 46, 47). In this model, the generalist and pulmonologist form a team that requires a good relationship, with open and effective communication with each other and with home care and nursing agencies, medical equipment companies, and all other

services involved. Delineation of who is responsible for what aspects of care, and communication of the specifics to all involved, is critically important. Families need to know who to call for each aspect of the child's care. For example, the pulmonology team would be accessed for help with acute respiratory problems and have a prominent, if not coordinating, role for hospital admissions. The generalist and team would be responsible for all aspects of primary care. For aspects of care that are not respiratory or primary care, the comanagement team would decide who would be responsible. For example, the pulmonology team may take responsibility for interacting with the dysphagia specialist and overseeing feeding management, due to the risk for aspiration. The generalist may assume oversight of the child's gastrointestinal, neurological, rehabilitation, and other care. Both components of the team should be attuned to integration of the child's medical care plan in nonmedical settings outside the home, notably in school. Direct communication and engagement of school staff on an enhanced level may be needed if more complicated elements of the child's care are to be delivered in that setting (i.e., airway clearance sessions, monitoring during any weaning process, behavioral interventions for a medical issue). It is also incumbent on the medical provider to review and adapt a specific plan able to accommodate advancement in other areas of the child's development (altering medication schedules or ventilator weaning for rehabilitative therapies sessions).

Families often need advice and support as they work to attain other, nonmedical services for their children. These will likely include optimum insurance coverage; transportation; and educational, vocational, and recreational services. A family, as a group or for an individual member, may also need support for their own optimal functioning (i.e., recognizing and treating stress or depression associated with care demands). Ideally, the child's care team includes one or more members adept at identifying, or better yet anticipating, these needs, with the knowledge and skills to help.

#### **Recommendation.**

1. For children requiring chronic home invasive ventilation, we suggest a comprehensive Medical Home

comanaged by a generalist and respiratory subspecialist (Strength of Recommendation: Conditional; Quality of Evidence: Very Low). The features of this Medical Home are summarized in Table 4.

### **Question 2: Should Standardized Discharge Criteria Be Used When Planning the Hospital Discharge of Children Requiring Chronic Home Invasive Ventilation?**

**Background.** Care in the home for children requiring chronic invasive ventilation is possible and preferred to care in an institution. Yet discharging a patient dependent on invasive ventilation can take many months or even years to arrange (48, 49). The decision to pursue chronic mechanical ventilation in the home setting will be influenced by the comfort level of the family and medical team caring for the child and the availability of resources in the child's home and community. It should be noted that many of the risks of chronic ventilation in children are present regardless of whether the child is cared for at home, in a long-term facility, or in the hospital setting (50, 51). The heterogeneity of insurance coverage and community availability of professional home care providers for necessary services and equipment makes comprehensive guidance difficult. The best judgment of the provider, after candid discussion with caregivers and review of available resources outside of the hospital, is essential for designing a customized discharge plan for each child.

The Workgroup found no published experimental or observational studies examining the use of standardized discharge criteria for children on chronic home mechanical ventilation. The effect of applying standardized discharge criteria on important outcomes in these children, such as death, rehospitalization, and quality of life, is not known. Guidelines designed to facilitate the safe discharge and home care of mechanically ventilated children were provided by the ATS in 1990, and subsequently pediatric guidelines have been published in the United Kingdom and Australia (52–54). The Pediatric Chronic Home Ventilation Workgroup compared existing discharge criteria from the chronic ventilation programs of seven children's hospitals within the United States and a composite document was generated. The Workgroup

reviewed the composite document and compared it to other consensus statements addressing pediatric home invasive ventilation in an effort to develop generalizable criteria for an ideal discharge (53, 54). When appropriate, aspects of care from adult guidelines for home mechanical ventilation were incorporated into the recommendations (55–57). On the basis of the extensive clinical experience of the Workgroup, we suggest the use of standardized discharge criteria to objectively assess readiness of an invasively ventilated pediatric patient for care in the home. The possible benefit of standardized discharge criteria would be to identify and eliminate important barriers to care in the home before discharge and consider alternate care arrangements if obstacles cannot be eliminated. Weight given to each component of the proposed criteria would vary between centers and patients. The recommendation places high value on the potential benefits of considering all facets of a child's care in the home before discharge and low value to the increased provider time and resource use that may be required.

Table 5 lists proposed standard criteria for discharge home from the hospital of children with invasive mechanical ventilation. All required services must be covered by insurance or other defined means, and a home care supplier must be identified to provide the necessary durable medical equipment (DME). In Table 5, section 1 describes the readiness of the child. Assessing the child's medical stability includes evaluating respiratory stability; ideally, ventilator settings have not required adjustment for weeks before discharge (54). Thresholds for delivered oxygen concentration or prescribed ventilator settings are often set by local home care company limitations and should be defined before discharge. Evaluation of home oxygen delivery systems should be performed to ensure the  $FiO_2$  delivered by home equipment is sufficient to meet the child's needs. Section 2 addresses the family caregivers. Families must have the ability to provide for the child's basic needs (safety, food, clothing, stimulation) while also managing the complex medical needs of their child. Caregivers should demonstrate competence with the provision of all therapies required by the patient before discharge. The Workgroup agreed that a minimum of two family caregivers should be trained in the care of the complex child

at home. One caregiver (including available professionals) should be awake and alert to respond to emergencies at all times. The requirement for number of caregivers in the home and the training of these caregivers is discussed in the following section of this document. Section 3 describes the basic requirements for a DME company, which will be needed to partner with the families and their health care team. In-home professionals are required for assistance to the family caregivers and, as described in section 4 and later in this document, must be appropriately trained. These professionals might include nurses, respiratory therapists, or other home personnel specifically trained in the care and management of the ventilated child. Section 5 addresses the important aspects of the home environment. The home environment must meet basic safety requirements and provide provisions for emergencies.

Advance care planning, including the development of written care directives, should be addressed before the child's initial hospital discharge. Anticipatory guidance that includes information about the patient's diagnosis and prognosis, potential emergency situations that may be faced, and the expectations for and limitations of available therapies should be provided. Decisions regarding resuscitation and life-sustaining treatments should be discussed with the family and, when appropriate, the patient. Patients and families should also understand that directives and goals of care can be modified at any time.

#### **Recommendation.**

2. For children requiring chronic invasive ventilation, we suggest the use of standardized discharge criteria to objectively assess readiness for care in the home (Strength of Recommendation: Conditional; Quality of Evidence: Very Low). Suggested components of these criteria are listed in Table 5.

### **Question 3: Should Home Caregivers Be Specifically Trained in the Care of Children Requiring Chronic Home Invasive Ventilation?**

**Background.** Children on home invasive ventilation are typically cared for by a

**Table 5.** Proposed Standardized Criteria for Discharge of an Invasively Ventilated Child to Home**1. The child must be medically stable for discharge.**

- No significant change to ventilator settings or oxygen requirement for at least several days and preferably several weeks before discharge.
- No acute decompensation events (e.g., PICU transfers) within the few days to weeks before discharge.
- Ventilator and oxygen requirements compatible with long-term medical stability and equipment available for home setting.
- Home respiratory equipment trialed and tolerated in the hospital for at least 24–48 h before discharge.
- Must tolerate the transport to and from hospital.

**2. Family caregivers must demonstrate the willingness and ability to care for the patient.**

- Caregivers must demonstrate competency in delivering all prescribed therapies (e.g., medication administration, feeding, respiratory care, CPR, home ventilator use, responding to monitors).
- Caregivers must demonstrate competency in the care and replacement of their child's tracheostomy, and caregiver education must include recognizing and responding to urgent issues such as tube obstruction, decannulation, and bleeding from tracheostomy.
- At least two family caregivers must be fully trained in all aspects of the child's care.
- Caregivers must understand the importance of the continual presence of an alert caregiver who can respond to alarms and emergencies.
- Caregivers must agree to care for their child in situations when additional services (such as in-home nursing) are not available even for extended periods of time.
- Caregivers should complete an independent stay before hospital discharge during which they are responsible for all aspects of the child's care (including responding to simulated emergencies).
- Routine hand washing is essential and its importance cannot be overemphasized.
- Caregivers must be able to safely transport the child in both routine and urgent situations (a "Go Bag" with all necessary travel items, including an extra tracheostomy tube and obturator, a size smaller tracheostomy tube, suction catheters, scissors, tracheostomy tube ties, and lubricant, will remain with the child at all times; disability parking privileges should be considered).
- Family caregivers should understand that if the child improves and no longer requires the same amount of professional caregiver support, they will be required to assume increasing responsibility for the child's care.
- Family caregivers must be instructed not to engage in cigarette smoking near the child and respiratory equipment, and smoking cessation should be encouraged.

**3. A DME company must be available and able to provide the required equipment and technical support.**

- The DME (or trained personnel from discharge facility) must perform a home inspection to confirm that the home environment and electrical systems are adequate for the necessary medical equipment.
- The DME company must provide 24-h availability as a resource and to service the equipment, including same-day replacement of malfunctioning equipment.
- DME respiratory clinicians should visit patients at least monthly and more often as needed.

**4. Professional in-home caregivers (e.g., nurses) as required to support the family must be arranged before discharge.**

- Home professional caregivers must maintain infant/child CPR certification.
- Professional caregivers must be required to achieve the competencies expected of the child's family-based caregivers.
- Each professional caregiver must complete ventilator training involving the specific type of ventilator used in the child's home.
- Professional caregivers must be available to meet the child at home on the day of discharge.
- An accredited agency must provide professional caregivers with experience in home mechanical ventilation and will maintain training to ensure maintenance of skills.
- Professional caregivers must be instructed not engage in cigarette smoking while on duty.

**5. The home and community environment must be safe and allow access to routine and urgent care as needed.**

- Primary care, pulmonary subspecialty care, and care coordination must be provided in a collaborative manner consistent with the family-centered care and Medical Home models.
- A formal safety plan should be posted near the patient to include: emergency contact numbers (EMS, primary care provider, specialty providers, DME contact, nursing agency) and any medical information essential to the child's care (allergies, medications, ventilator settings, specific instructions).
- A functioning phone must remain with the patient in case of emergency.
- The home should be safe and free from fire/health/safety hazards and provide easy access to the child at all times.
- The home must have a functional fire extinguisher that home occupants are able to operate. A home fire escape plan that includes the patient and minimal equipment needed for life support should be in place.
- The ambient temperature in the home should remain within the range recommended by the ventilator's manufacturer.
- Irritants (e.g., cigarette smoke, incense burning, molds) should not be present.
- Local EMS should be made aware of the patient and the patient's condition. On the basis of distance from emergency services, consideration should be given to additional back-up equipment in home.
- Letters requesting that services be restored quickly in an outage should be sent to the telephone and utility companies.

*Definition of abbreviations:* CPR = cardiopulmonary resuscitation; DME = durable medical equipment; EMS = emergency medical/transport services; PICU = pediatric intensive care unit.

combination of family and professional caregivers. In the ideal situation, the child's caregivers understand the patient's disease and the role invasive ventilation plays in its management, the mechanics of providing invasive ventilation including the features

and limitations of the specific ventilator used, and tracheostomy care with a focus on preventing tracheostomy-specific complications. Caregivers must provide comprehensive care in all eventualities, particularly in emergencies. Accidental

deaths remain a significant cause of morbidity in this population, and hospital readmissions are common. During the past three decades, the overall mortality rate of a total of 1,008 children enrolled in the Pennsylvania Ventilator Assisted Children's

Home Program (VACHP) was 27.5%. Despite technological advances in home monitoring, 18 to 20% of the overall mortality rate was attributed to preventable deaths related to tracheostomy accidents that included undetected decannulation and mucous plugging of the tracheostomy tube (58). In a retrospective observational cohort analysis of 228 children enrolled in a home mechanical ventilation program in southern California over a 22-year period (990 person-years), there was a 21% mortality rate. Progression of underlying conditions accounted for only 34% of the deaths, whereas 49% of the deaths were unexpected. Of the unexpected deaths, 19% were related to airway issues, including acute airway obstruction, tracheal hemorrhage, and accidental decannulation (51). A study from the same program in California found that 40% of newly discharged pediatric patients requiring home mechanical ventilation required nonelective readmission to the hospital in their first year home; half of these admissions occurred in the first 3 months (59). Pneumonia and tracheitis were the most common causes of readmission, and the authors postulate that this may be, in part, related to inadequate caregiver training on airway clearance and care. A study from Germany examining severe emergencies in a group of ventilator-dependent children found an incidence of 0.27 emergencies per patient-year and also found that the majority were related to respiratory problems, such as infection, atelectasis, and increased secretions and tracheostomy complications (60).

Authors reviewing outcomes in this population have suggested that improved caregiver training may help reduce morbidity and mortality on the basis of their clinical experience (58, 59). Consensus statements by the ATS and the American Academy of Otolaryngology–Head and Neck Surgery that describe the care of children with tracheostomies emphasize the importance of home caregiver education, which includes both family members and home nurses (61, 62). The Workgroup found no experimental or observational studies directly addressing the question of the effect of training caregivers or the presence of a trained caregiver on important outcomes, such as death or recurrent hospitalization, for children requiring chronic home invasive

ventilation. Despite the very low quality evidence available, the Workgroup, on the basis of consensus opinion and unsystematic observations, strongly recommends that an awake and alert trained caregiver be always present and immediately available in the home of a child requiring chronic invasive ventilation.

In making this strong recommendation, the Workgroup believes that the desirable consequences would clearly outweigh the undesirable consequences, which include increased use of resources and a possible disruption to families, of following this recommendation. Lack of an awake and attentive trained caregiver would place the child in a life-threatening situation. Training is irrelevant if a caregiver cannot respond to an urgent situation secondary to sleepiness or fatigue. To provide an awake and alert trained caregiver at all times generally requires the involvement of both professional and nonprofessional (family) caregivers.

Previous guidelines released by the ATS on the care of a child with a tracheostomy in the home recommended that two adult caregivers be trained in the care and replacement of a tracheostomy tube (61). A small retrospective study surveyed caregivers of infants discharged from the hospital with a tracheostomy. Most caregivers (70%) reported that the second caregiver participated in the child's medical care often or very often (63). Fifty percent of the respondents believed they could not have cared for the infant in the home without another trained caregiver. On the basis of the experience of the Workgroup and these limited survey data, we suggest that at least two family caregivers are trained to provide care for the child in the home. This recommendation places a high value on the safety of the patient and the quality of life of the caregivers and low value in the increased resource use required for training more than one caregiver.

Limited literature describing training of both family and professional caregivers was available and is reviewed below. Data examining to what extent level of training affects the outcomes of these patients are not available.

**Family caregivers.** Single-center observational studies have outlined recommended training for caregivers

(64–68). Recommendations include a comprehensive and staged educational approach that begins even before placement of the tracheostomy and spans until the time of the child's discharge from the hospital. These approaches have applied various aids that involve individualized instruction, audiovisual and written materials, supervised practice using home equipment, and use of dolls or mannequins for skill demonstration. Programs have included written objectives and validation of acquired skills and knowledge. Structured educational programs have been shown to reduce the length of the initial hospital stay after tracheostomy placement in children (69, 70). Training is designed to ensure parental familiarity and understanding of the role and use of devices and equipment; comfort with routine tracheostomy and ventilator care, day-to-day respiratory management, and troubleshooting; as well as emergency care. A study by Kun and colleagues demonstrated, through the use of a 25-question survey, that family caregivers once trained have a good understanding of how to respond to ventilator or tracheostomy emergencies, with no difference in scores noted between lay caregivers and licensed nurses (71). Some deficits in knowledge were found, however, with the authors recommending more in-depth education on the limitations of ventilator alarms.

**Professional caregivers.** Care of a chronically ventilated child in the home can rarely be accomplished without the use of professional caregivers. As noted previously, safe care of a child with a tracheostomy and home mechanical ventilator relies not only on caregiver training but also on the presence of an awake and alert trained caregiver. Professional caregivers are commonly used to meet this standard and provide the needed respite for families to sleep, work, and attempt to maintain life balance. In a survey of families caring for ventilator-dependent children, 25% reported providing direct patient care to their child for more than 16 h/d, and 40% reported an unmet need for skilled nursing in the home (72). A significant portion of family caregiver respondents reported frequent depressive symptoms, and, in this cohort, screening positive for a likely depressive disorder was associated with reporting unmet needs for care. In

another small study of caregivers of ventilator-dependent children, symptoms of clinical depression and sleep deprivation were significantly associated with a lack of nighttime nursing (73). Guidelines addressing the use of professional home nursing care for children with a tracheostomy or tracheostomy with ventilator have stressed the use of home nursing care during the transition from hospital to home and supported that many children will have an ongoing need for home professional care (53, 61). The number of hours of professional care required will be based on multiple factors (for example, complexity of the child's care, the child's dependence on the ventilator, the family's ability to provide safe care, and the other demands on the family's time, including other children) and must be evaluated and reevaluated as situations change on a case-by-case basis.

In Pennsylvania, a care assessment survey was administered to 107 randomly selected families enrolled in VACHP. These families reported that home care was frequently disrupted by 1) the lack of available nurses, and 2) an inadequate level of skill demonstrated by the homecare nurses (74). In response to the survey results, the organization assessed the skill levels of 21 experienced home care nurses (16 registered nurses and 5 licensed practical nurses) who were providing care to the VACHP children. The assessments revealed a knowledge deficit in many areas of care. The deficient areas were: respiratory assessment (71%), tracheostomy care (43%), tracheostomy string change (90%), suctioning of the tracheostomy (95%), tracheostomy tube change (90%), tracheostomy emergencies (86%), cardiopulmonary resuscitation with a tracheostomy (47%), ventilator care and management (86%), and troubleshooting ventilator alarms (100%). Individual training included pretraining evaluation, individual skill level validation, simulation, return demonstration, and post-training evaluation to all the nurses over a 7- to 13-hour period. At the conclusion, each nurse was able to demonstrate mastery of the skills with the patient. Parents reported an overall increase in satisfaction with the care provided by their nurses post-training (74).

From 2003 to 2012, VACHP provided training to 600 home care nurses from across the United States caring for

ventilator-assisted children. Of the nurses who identified themselves as experienced in tracheostomy and ventilator home care, the average pretest score was only 60%, with some nurses scoring as low as 20%. Post-training evaluation was conducted with each of the nurses to assure skill acquisition to provide proficient care in the home (58). Kun and colleagues used home mechanical ventilator emergency scenarios provided by an online survey to evaluate the knowledge of practicing pediatric home health nurses (75). Regardless of years of experience, the nurses scored poorly, and those surveyed overwhelmingly favored more training in home mechanical ventilation. The authors recommend enhancing nurse education through the use of simulation, "teach back" methods, and return demonstrations and simulations in the home where the nurse is providing care. It is unknown, however, whether periodic assessment and reeducation of professional caregivers reduces the frequency of adverse events in the home.

On the basis of the literature reviewed and the unsystematic observations of the Workgroup, we suggest that ongoing education to allow acquisition of and then reinforce and augment the skills required for patient care be provided to both family and professional caregivers of children requiring chronic home invasive ventilation. Ongoing education after the initial training period would help reinforce learned skills and allow training on new protocols and technology. Education should be provided by the involved medical professionals in the clinic or hospital, professional personnel agencies, and DME companies. This recommendation places a high value on safety and the potential clinical benefits to the patient and a low value on potential increased cost and resource use.

#### **Paraprofessional caregivers.**

Nonprofessional caregivers are rarely employed in the home care of chronically ventilated children in the United States, and research literature is limited. An Australian study of 168 ventilator-dependent children at home evaluated care provided by alternative caregivers in place of skilled nurses (76). The study reported that the trained lay caregivers provided adequate care and did not correlate negative patient outcomes with care provided by nonprofessionals. However, the study did not describe the training curriculum or methodology. Most

of the children in the study required noninvasive ventilation; therefore, further investigation of the use of paraprofessional caregivers for children who require conventional mechanical ventilator support is warranted.

#### **Recommendation.**

- 3a. We recommend that an awake and attentive trained caregiver be in the home of a child requiring chronic invasive ventilation at all times (Strength of Recommendation: Strong; Quality of Evidence: Very Low).
- 3b. For children requiring chronic invasive ventilation, we suggest that at least two specifically trained family caregivers are prepared to care for the child in the home (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
- 3c. We suggest that ongoing education to acquire, reinforce, and augment skills required for patient care be provided to both the family and professional caregivers of children requiring chronic home invasive ventilation (Strength of Recommendation: Conditional; Quality of Evidence: Very Low). Suggested educational objectives are provided in Table 6.

#### **Question 4: Should Standard Home Equipment Requirements Be Applied When Planning for the Equipment Needs of Children Requiring Invasive Ventilator Support in the Home?**

**Background.** The Workgroup found no experimental or observational studies directly addressing the question of whether the availability of certain equipment in the home of a child dependent on invasive ventilation would lead to improved outcomes and therefore be considered a required component of care. We found several studies examining the performance of equipment in the home and one evaluating the performance of ventilator alarms in a setting designed to simulate accidental decannulation, and these are described in the following paragraphs. None of the literature reviewed addressed the effect of providing specific home equipment or of maintaining home equipment on outcomes such as death, morbidity, or quality of life. The indirect evidence described below,

**Table 6.** Suggested Educational Objectives for Family and Professional Caregivers of Children Requiring Home Mechanical Ventilation**Pulmonary care and assessment**

Caregivers should be able to:

- Obtain basic vital signs, including pulse rate, breath rate, and oxygen saturation
- Recognize and state the child's usual breath rate, pulse rate, oxygen saturation, respiratory effort, and color
- Identify type of ventilator used in home and ventilator settings
- Identify type of tracheostomy tube and whether it is cuffed or uncuffed and, if cuffed, instructions for inflation of the cuff (manufacturer recommendation for cuff inflation [air vs. water], amount of air/water used to inflate cuff, hours per day spent with cuff inflated)
- Identify type of "back-up" tracheostomy tube to be used if primary tube cannot be replaced
- Know that an additional tracheostomy tube, the back-up tracheostomy tube, and the supplies needed to change the tracheostomy should be with the child at all times (the "Go Bag")
- Identify signs of respiratory distress and describe proper intervention. (Signs of respiratory distress to review may include: increased tracheal secretions or change in color of secretions, retractions, increased work of breathing, cough, color changes, nasal flaring, increased or decreased heart rate, increased or decreased respiratory rate, desaturation, anxiety, abnormal breath sounds.)

**Tracheostomy care**

Guidelines for tracheostomy education are provided in Reference 61

**Emergency response**

Caregivers should be able to:

- Verbalize criteria for calling emergency services (such as 911 in the United States)
- Be certified in CPR
- Be able to access a list of numbers for emergencies or problems not requiring emergency services (physicians, DME, etc.) and know who to contact on the basis of the type of emergency
- Demonstrate use of self-inflating bag and mask in routine and emergency care procedures
- List signs of tracheostomy obstruction
- Demonstrate appropriate suctioning techniques to remove tracheostomy obstruction
- Demonstrate an emergency tracheostomy tube change (change done by one caregiver without assistance)
- Demonstrate knowledge of emergency medications (if applicable)
- Verbalize plans for loss of electricity, fire, tornado, or other natural disaster

**Ventilator training**

Caregivers should be able to:

- Identify electrical power sources
- Assemble ventilator circuit and humidification system
- Describe routine cleaning of equipment
- Add oxygen to circuit if indicated
- Verbalize that ventilator alarms must be audible throughout the home
- Demonstrate how to properly turn the ventilator on, test the ventilator before use, and view and verify settings
- Demonstrate an understanding of ventilator alarms and how to troubleshoot the alarms
- Demonstrate the appropriate technique for draining tubing in the ventilator circuit, down and away from child
- Demonstrate the ability to keep battery-operated back-up equipment charged and ready for use
- Demonstrate how to charge batteries for ventilators
- Demonstrate how to connect and use the external battery for the ventilator if applicable
- Verbalize understanding of the approximate battery life for each piece of equipment

**Infection control practices**

Caregivers should be able to:

- Demonstrate infection control practices as they relate to the plan of care
- Demonstrate proper hand-washing technique
- Demonstrate proper disposal of contaminated material
- Demonstrate how to clean and disinfect reusable medical supplies

**Medications**

Caregivers should be able to:

- Identify the dosage and frequency of all medications required by the child
- Explain the indications and side effects of medications
- Demonstrate the ability to prepare and administer medications correctly

**Oxygen**

Caregivers should be able to:

- Demonstrate the proper care and use of home oxygen delivery equipment (e.g., cylinders, concentrators)
- Verbalize the safety issues related to use of oxygen in the home (risk of fire with smoking, open flames, flammable products near the oxygen, or close proximity to heat sources)
- Demonstrate understanding of when and how oxygen should be used for the patient

**Oximetry monitoring**

Caregivers should be able to:

- Demonstrate the proper and secure placement of the oximeter probe
- Demonstrate the ability to differentiate true from false oximeter readings
- Verbalize steps for responding to an oximeter alarm
- Verbalize an understanding of normal oxygen saturations and a plan for responding to saturations that fall below the normal range

(Continued)

**Table 6.** (Continued)**Suctioning equipment**

Caregivers should be able to:

- Demonstrate the correct catheter size to use for the patient
- Demonstrate the correct suction pressure and catheter depth to use to clear tracheostomy
- Demonstrate how to test for suction pressure
- Demonstrate a clean suction technique

**Nebulizer/metered dose inhaler (if ordered)**

Caregivers should be able to:

- Verbalize when the prescribed inhaled medications are required
- Demonstrate how to deliver inhaled medications in ventilator circuit if applicable

*Definition of abbreviations:* CPR = cardiopulmonary resuscitation; DME = durable medical equipment.

Further instruction and competency assessment will be required if patient requires equipment/therapy beyond those listed above (e.g., pulmonary clearance therapies, cardiorespiratory monitors, end-tidal CO<sub>2</sub> monitors, etc.). The goal should be to ensure that all caregivers are fully versed in all aspects of the child's care.

coupled with the unsystematic observations of the Workgroup experts, was used to formulate the recommendations.

Children requiring invasive home ventilation have diverse underlying etiologies for their chronic respiratory failure and often have significant associated comorbidities. They are at high risk of death and require significant medical and technological support in the community setting. Although most of the deaths seen in patients requiring chronic home invasive ventilation are related to the progression of their underlying disease or their associated comorbidities, a significant number of these deaths are caused by tracheostomy-related accidents or complications, many of which may be preventable (51).

Although many of these tracheostomy-related deaths may be due to problems regarding caregiver training and their ability to appropriately respond to emergency situations, there may be equipment-based strategies available to avoid or reduce tracheostomy-related mortality. One means may be through using appropriate monitoring strategies in the home. Although positive-pressure ventilators are equipped with various alarms, these may not provide adequate protection. A study by Kun and colleagues found that low inspiratory pressure alarms failed to alarm during simulated decannulation when tested with the smaller-diameter tracheostomy tubes often used in pediatric patients (77). This suggests the need for alternate monitoring strategies. In a separate study examining the performance of ventilators in the home environment, poor functioning of ventilator alarms was also noted, with 18.6% of the built-in alarms for disconnection and 5.1%

of the alarms for obstruction in the home ventilators found to be nonfunctional when investigated by the examiners (78). Limited available data suggest that pulse oximetry is the preferred monitoring method, although this is not provided for every patient, as reported in a 2007 study of home ventilated patients in Massachusetts (22). Standard home cardiorespiratory monitors that monitor heart rate and chest wall movement will only alert caregivers if there is absence of respiratory effort and/or associated bradycardia. In infants and children, hypoxemia is likely to be an early indicator of airway obstruction or equipment malfunction leading to inadequate ventilation, whereas bradycardia and central apnea are later complications of a serious event, which make patient resuscitation more difficult. For children requiring chronic home ventilation, we suggest monitoring, especially when the child is asleep or unobserved, with a pulse oximeter. In making this recommendation, we place high value on the potential benefits of monitoring with a pulse oximeter and low value on the potential risks, which may include false alarms (which can lead to caregiver desensitization) as well as difficulty maintaining the pulse oximeter probe in place.

Unlike tracheostomy-related issues, deaths secondary to ventilator failure are rare. Equipment malfunction and/or failure, however, can lead to significant overuse of resources by patients. Equipment failure may be related to age of the equipment, longer intervals between equipment servicing, longer ventilator use by the patient, failure of caregivers to care for

equipment appropriately, and failure of the caregivers to recognize poor equipment performance (e.g., leaks in the circuit) (79). Interestingly, equipment failure was also associated with "very new" ventilators, suggesting issues are uncovered after release of newer models (79). Strategies for addressing potential equipment failure include regular maintenance and servicing of home equipment as well as regular reeducation of the patients and caregivers. A study by Farre and colleagues examining quality-control procedures in Europe found that medical centers prescribing home mechanical ventilation were generally not involved in the care and maintenance of the ventilator and that ventilator companies were servicing equipment with a frequency that ranged from 3 to 12 months (80). Most prescribing centers were unaware of the quality-control procedures followed by the ventilator companies, with communication between the two entities limited. The provision of appropriate back-up supplies was found to decrease the risk of ventilator malfunction and associated mortality. A 2011 German study of 54 children and adolescents receiving ventilation in the home setting reported only three acute emergencies associated with ventilator failure without resulting mortality, subsequent hospitalization, or persistent adverse event (60). Authors of this study postulated that the low incidence of adverse events associated with equipment malfunction in their patient population may have been related to the provision of back-up ventilators, self-inflating bag and masks, and emergency oxygen for all of the ventilator-dependent patients. Providers in the United States should be aware of the

requirement to report medical device failures or defects to the Federal Drug Administration (81).

Even in the absence of ventilator malfunction or failure, patients may be at risk for underventilation or overventilation in the home setting. Studies in adult patients have suggested that performance of ventilators in the home may vary considerably and that there is often a difference between the set and the prescribed ventilator settings on these ventilators. This places patients at risk for hypoventilation and associated complications (78). In addition, in transitioning a patient from an internal positive end-expiratory pressure ventilator to an external positive end-expiratory pressure ventilator (or vice versa), the clinician must recognize that the required settings on the ventilator may be different so as to achieve the same ventilatory support. Home ventilators have been shown to vary in their ability to reliably deliver a specified tidal volume and compensate for air leaks (82, 83). Pediatric patients should demonstrate stability on their chosen home device before discharge and may require readmission or increased monitoring in the home if transition from one home ventilator type to another is required.

For children requiring invasive home ventilation, we recommend regular maintenance of ventilators and associated equipment in the home as outlined by the manufacturer. Care should be taken to assure that the actual ventilator settings as seen on the control panel match the prescribed settings. Twenty-four hour a day service and phone support for mechanical failure or malfunctions must be available and is generally provided by the DME company. This recommendation supports the safe and appropriate use of all equipment chosen by the provider for use in the home.

A 1998 consensus statement from the United Kingdom recommended that equipment chosen for the home for children requiring long-term ventilation be “portable, durable and simple to use” (53). A second ventilator was believed to be required for any child who cannot be off the ventilator for 6 continuous hours. It was also recommended that alternative power sources be supplied (battery or generator). Last, it was recommended that clear plans should be in place for the maintenance and

replacement of equipment in the case of mechanical failure. Given the limited data available to the clinician when choosing appropriate equipment for the home and the absence of any randomized controlled trials, our expert panel was surveyed to formulate equipment recommendations. Each pediatric tertiary center with representatives in the working group was asked to provide their written protocol for standard equipment prescribed at initial discharge for home ventilated infants or children. These lists were then merged into a single list containing only DME; disposable items were not included. The final list included 21 items. All participants were then e-mailed a survey asking them to 1) rank-order the 21 items from most important to least important; and 2) rate whether each item was absolutely required, required, strongly recommended but not required, or optional. A total of 15 experts in chronic home ventilation from 15 different centers responded to the survey. Participants were encouraged to discuss the survey with their colleagues and/or hospital discharge planner. Survey results are summarized in Table 7. Equipment believed to be absolutely required by 70% or more of the experts surveyed included: the ventilator, a back-up ventilator, batteries for the ventilator, self-inflating bag and mask, suctioning equipment (portable), heated humidifier, supplemental oxygen for emergency use, nebulizer, and a pulse oximeter (nonrecording). The remainder of the survey results can be found in Table 7. We suggest that the “absolutely required” equipment identified in our survey of expert clinicians be available in the home. On the basis of experience, the Workgroup believed the presence of specific pieces of equipment could prevent the development of life-threatening situations and/or reduce their severity. This recommendation places high value on the potential to avoid emergent situations due to the presence of important reserve and emergency equipment and low value to increased resource use and increased equipment costs.

Another possible strategy to avoid complications for patients dependent on invasive ventilation is to decrease the risk of tracheostomy plugging through the optimization of airway clearance. The recent Canadian Thoracic Society statement on home ventilation, which

focused on the adult population, strongly recommended manually assisted cough techniques or the use of the cough assist in patients with tracheostomy, regardless of the underlying etiology of the chronic respiratory failure (57). Methods for augmenting cough, including MI-E devices, are strongly recommended for use in patients with neuromuscular disease with an ineffective cough so as to help maintain airway patency and avoid respiratory complications, such as atelectasis and infection (84–86). We suggest that an MI-E device be used to help maintain airway patency in patients requiring home mechanical ventilation with ineffective cough, including, but not limited to, those with neuromuscular disease with poor respiratory muscle strength. This recommendation places high value on the potential to avoid airway plugging and low value on increased costs and resource use.

#### *Recommendation.*

- 4a. For children requiring chronic home invasive ventilation, we suggest monitoring, especially when the child is asleep or unobserved, with a pulse oximeter rather than use of a cardiorespiratory monitor or sole use of the ventilator alarms (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
- 4b. For children requiring chronic home invasive ventilation, we recommend regular maintenance of home ventilators and all associated equipment as outlined by the manufacturer.
- 4c. We suggest the following pieces of equipment for use in the home when caring for a patient on home mechanical ventilation: the ventilator, a back-up ventilator, batteries, a self-inflating bag and mask, suctioning equipment (portable), heated humidifier, supplemental oxygen for emergency use, nebulizer, and a pulse oximeter (nonrecording) (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).
- 4d. We suggest that an MI-E device be used to help maintain airway patency in patients requiring home mechanical ventilation with ineffective cough, including, but not limited to, those with neuromuscular

**Table 7.** Survey of Expert Opinion on the Necessary Equipment for Care of Children Requiring Invasive Home Ventilation

Equipment	“Absolutely Required” or “Required” (% of Experts)	“Strongly Recommended” (% of Experts)	“Optional” (% of Experts)	No. of Respondents*
Battery (long)	100.0	0	0	15
Heated humidifier	100.0	0	0	15
Suction (portable)	100.0	0	0	15
Ventilator (primary)	100.0	0	0	14
Oxygen	93.3	6.7	0	15
Pulse oximeter (nonrecording)	93.3	6.7	0	15
Self-inflating bag and mask	93.3	6.7	0	15
Battery (short)	78.6	7.1	14.3	14
Nebulizer	73.3	20.0	6.7	15
Ventilator (back-up)	71.4	28.6	0	14
Suction (stationary)	64.3	28.6	7.1	14
Stethoscope	53.3	33.3	13.3	15
Mechanical insufflation–exsufflation device	33.3	26.7	40.0	15
Car adapter	26.7	73.3	0	15
Cardiorespiratory monitor (recording)	21.4	28.6	50.0	14
Pulse oximeter (recording)	21.4	42.9	35.7	14
Test lung	21.4	21.4	57.1	14
Cardiorespiratory monitor (nonrecording)	13.3	26.7	60.0	15
Tool chest	7.7	46.2	46.2	13
Capnograph	7.1	35.7	57.1	14

\*Pediatric tertiary care centers surveyed include: Arkansas Children’s Hospital (AR), Lucille Packard Children’s Hospital at Stanford (CA), Children’s Hospital Colorado (CO), All Children’s Hospital (FL), Rush Children’s Hospital (IL), The University of Chicago Medicine Comer Children’s Hospital (IL), Riley Hospital for Children (IN), Johns Hopkins Children’s Center (MD), St. Louis Children’s Hospital (MO), Duke Children’s Hospital & Health Center (NC), Stony Brook Children’s Hospital (NY), Nationwide Children’s Hospital (OH), Children’s Hospital of Pittsburgh (PA), Ventilator Assisted Children’s Home Program (PA), and Hasbro Children’s Hospital (RI).

disease with poor respiratory muscle strength (Strength of Recommendation: Conditional; Quality of Evidence: Very Low).

**Research Priorities**

These guidelines are based largely on the extensive clinical experience of the Workgroup members. Published data are limited and experimental, and even observational clinical studies are absent. The following future directions were identified by the Workgroup in hopes that study of this population will lead to improved care. We anticipate that updates to these guidelines will be required as our knowledge increases and home equipment changes. We recognize that what works in one center may not work well in another. Regular review and revision of these guidelines with individual patients and their community partners (primary care providers, home nursing agencies, DME companies, emergency medical services) is needed to ensure the guidelines remain relevant in the local setting.

1. A national registry of patients on home invasive ventilation should be established to monitor the epidemiology of children requiring chronic invasive ventilation in the home, track outcomes, and facilitate multicenter research. A learning network associated with the registry could be developed and regularly bring together stakeholders to share knowledge and develop and implement research projects and clinical programs.
2. A national safety reporting system of severe events in the home that would enable tracking of poor outcomes or dangerous situations due to problems in the health care delivery system should be developed. Analysis of these events will help to alert caregivers to practices or equipment that place patients at risk and support important quality-assurance efforts, which can be used to tailor the guidelines. We also propose standardization of nomenclature for ventilator modes and settings to avoid errors leading to inappropriate settings or ventilator support. Proprietary naming of ventilator modes by manufacturers results in different terms being used for the same or similar ventilator modes and leads to confusion among medical staff.
3. Evaluation of the efficacy of formalized family caregiver training programs and professional caregiver training programs is needed. The role of simulation programs and other media should be investigated. To possibly expand the available home care workforce and provide needed support to families, research examining the role and required training of paraprofessional caregivers for children requiring chronic home invasive ventilation should be performed.
4. Studies examining the role of telemedicine and remote monitoring in the care of chronically ventilated children should be undertaken, specifically investigating the usefulness regarding patient outcomes and equipment troubleshooting.
5. Further research is needed on the most effective models of primary care–specialist partnerships. Research regarding the delivery of health care to

children dependent on long-term mechanical ventilation will require development of innovative models of health care efficiency that include metrics that reflect: quality; medical outcomes; overall health care cost, including both dollars paid and time

invested by all members of the health care team; and patient/family experience.

### Conclusions

There are many barriers to caring for the ventilator-dependent child outside of the

hospital, but practitioners, families, and patients see the benefits of care in a home setting daily. The Workgroup hopes that continued research and efforts to share experiences will lead to improved care and enhanced quality of life for these patients. ■

This official clinical practice guideline of the American Thoracic Society was prepared by an *ad hoc* subcommittee of the Assembly on Pediatrics.

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