Long-Term Home Mechanical Ventilation in the United States

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Summary

Patients requiring prolonged mechanical ventilation are rapidly increasing in number. Improved ICU care has resulted in many patients surviving acute respiratory failure to require prolonged mechanical ventilation during convalescence. Also, mechanical ventilation is increasingly used as a therapeutic option for patients with symptomatic chronic hypoventilation, with an increased effort to predict nocturnal hypoventilation to initiate ventilation earlier. There are continued efforts by ventilator manufacturers to improve home ventilators. These factors point to a likely increase in the number of patients receiving home mechanical ventilation in the United States. Unfortunately, there are no comprehensive databases or national registry of home ventilator patients—therefore the number of home ventilator patients is unknown. There are real challenges to providing mechanical ventilation in the home, which include caregiver training, adequacy of respiratory care, and reimbursement. *Key words: discharge; home; ventilation; safety; tracheostomy.* [Respir Care 2012;57(6): 921–930. © 2012 Daedalus Enterprises]

Introduction

Mechanical ventilation in the home is not a new idea. Patients with poliomyelitis benefited from home mechanical ventilation by iron lung during the years before the 1950s. With the development of positive pressure ventilators, home use of these devices with tracheostomies emerged as a viable technology. In 1977, the LP3 portable volume ventilator was approved by the FDA for use outside the hospital.[1] The initial target population for home mechanical ventilation was ventilator dependent pediatric patients. This effort was led by Dr Allen Goldberg, who learned of this approach in France and returned to Children's Hospital of Pennsylvania to begin the first home ventilator program there (personal communication, Allen Goldberg, 2012).

An important reimbursement milestone occurred in 1981 with Katie Beckett, an infant with viral encephalitis that left her ventilator-dependent in a pediatric ICU in Iowa. Officials from Medicaid, the government health insurance plan for the needy, initially claimed that it could

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only pay for her care in the pediatric ICU. At the behest of her family (and others) the Medicaid waiver program (sometimes referred to as the "Katie Beckett waiver") was passed. This program allowed parents like the Becketts, who made too much money to qualify for Medicaid, to be covered for their child with extreme medical costs, even though their child lived at home instead of in an institution.[2]

Technology, expertise, and funding were now available to support ventilator dependent patients outside of the hospital. The door was now open for many chronic ventilator patients, both children and adults, to live at home. The purpose of this paper is to review some of the history and research leading to current practices in home mechanical ventilation. This review will also address prevalence, outcomes, and barriers to successful implementation of home mechanical ventilation. Additionally, practical tips for the care of the patient receiving home mechanical ventilation can be found in the online supplement that accompanies this paper.

Note: Some people use the terms "ventilator-assisted individual" or "vent user," rather than "patient," as the preferred descriptor for a person who is chronically dependent on a mechanical ventilator.[3] These individuals consider the mechanical ventilator simply as an assistive device, similar to a walker or wheelchair—and therefore deem the term "patient" as inappropriate. For ease of reading, and considering that the primary audience for this paper is clinicians, the term "patient" will respectfully be used in this paper.

Why Is Home the Preferred Location for Long-Term Mechanical Ventilation?

Ideally, the preferred location for long-term mechanical ventilation is in the home, because costs are reduced (hospital costs $21,570, homecare costs $7,050, dollar savings per patient, per month $14,520),[4] quality of life is enhanced, and integration into the community is maximized.[5] For the pediatric ventilator patient, the advantages of home ventilation also include being reunited with parents and family, which greatly enhances normal development and relationships. Home mechanical ventilation also reduces exposure to hospital-borne infections, and frees hospital ICU beds for other acutely ill patients.[6] Table 1 lists some of the important differences, from the patient's perspective, between the hospital ICU and the patient's home.[7]

Table 1. Patient's View of the Environment, ICU Versus Home

ICU	Home
Noise	Relative quiet
Light	Day/night cycles
Limited view of the world	Outdoors easily visible and probably accessible
Crowded/cramped	Relatively roomy
Limited visitation allowed	Supportive visitors encouraged
Immobilized	Mobility increased
Sterile surroundings	Personal objects
Little control	More independence
Communication limited (or not a priority)	More time for speech development
High reliance on technology	More reliance on family supervision
Limited staff nurturing time	Family nurturing

(Adapted from Reference 7.)

Appropriate Patients for Home Mechanical Ventilation

The indications for mechanical ventilatory support in the home are increasing as technology and infrastructure support improve. Tables 2 and 3 summarize both adult and pediatric medical conditions suitable for home mechanical ventilation. However, the levels of support and the goals of support can differ substantially, depending upon a number of factors. For example, some conditions may require support only at night and/or intermittently during waking hours. Other conditions, however, require high levels of support 24 h/d. Disease trajectory can also affect goals and planning. For example, a slowly improving process (eg, the adult recovering from chronic critical illness) may warrant plans for weaning the support. On the other hand, progressive respiratory failure will need the incorporation of plans for future palliative care. Indeed, in some progressive diseases (eg, type 1 spinal muscle atrophy), the institution of tracheostomy and high level support is sometimes challenged as an inappropriate intervention in a rapidly fatal disease.[8]

The role of invasive versus noninvasive ventilation (NIV) in the home is frequently debated. Advantages to NIV relate to the fact that no artificial airway is required, and thus there is a reduced risk of tracheal damage (eg, tracheomalacia, tracheal erosion, tracheal rupture) and speech alterations. Artificial airways are also associated with discomfort, a moderate increase in the risk of infections, increased costs, and a higher burden of care. On the other hand, invasive ventilation via tracheostomy is associated with a reduced risk of aspiration, may be more comfortable when required for more than 20 hours per day, and may allow long-term survival for those patients choosing to continue life-long mechanical ventilator support.[5] In general, most agree that a requirement for only nocturnal support warrants NIV, while a requirement for 24 h/d support often warrants support through a tracheostomy. The decision on invasive versus NIV for patients needing more than simple nocturnal support but less than 24 h/d support must be individualized. Some clinicians argue that

Table 2. Medical Conditions That May Be Appropriate for Home Mechanical Ventilation

Central Nervous System Disorders
Arnold-Chiari malformation
Central nervous system trauma
Cerebrovascular disorders
Congenital and acquired central control of breathing disorders
Myelomeningocele
Spinal cord traumatic injuries
Neuromuscular Disorders
Amyotrophic lateral sclerosis
Guillain-Barré syndrome
Muscular dystrophies
Myasthenia gravis
Phrenic nerve paralysis
Polio and postpolio sequelae
Spinal muscle atrophy
Myotonic dystrophy
Skeletal Disorders
Kyphoscoliosis
Thoracic wall deformities
Thoracoplasty
Cardiovascular Disorders
Congenital and acquired heart disease
Upper Airway Disorders
Pierre-Robin syndrome
Tracheomalacia
Vocal cord paralysis
Lower Airway Disorders
Bronchopulmonary dysplasia
COPD
Cystic fibrosis
Complications of infectious pneumonias
Pulmonary fibrotic disease

(Adapted from Reference 3.)

Table 3. Pediatric Medical Conditions That May Be Appropriate for Home Mechanical Ventilation

Increased Respiratory Load
Bronchopulmonary dysplasia
Hypoxia and hypercapnia
Pediatric COPD
Restrictive parenchymal lung disease
Chest wall abnormalities
Ventilatory Muscle Weakness
Motor neuron disease
Primary myopathies (eg, Duchenne muscular dystrophy)
Spinal cord injury
Failure of Neurologic Control of Ventilation
Congenital central hypoventilation syndrome
Acquired central hypoventilation syndrome (eg, brainstem tumor, hemorrhage)
Myelomeningocele
Developmental disorders or neurologic control of breathing (eg, apnea of prematurity)

(Adapted from Reference 3.)

using a noninvasive interface 24 hours a day may be uncomfortable and cosmetically suboptimal and therefore a tracheostomy should be performed. Other clinicians point to evidence demonstrating that many individuals have been successfully ventilated for up to 24 h/d for many years with noninvasive methods—most often a mouthpiece during the day and a mask of some type at night.[9] Table 4 reviews the indications for home mechanical ventilation, both noninvasive and invasive.[3,5]

Table 5 reviews general criteria to determine if the patient is stable and ready for discharge to the home, as well as some potential contraindications for home mechanical ventilation.[10,11] Note that some of the concerns raised in Table 5 have been addressed in recent years. For example, one of the latest home ventilators offers an optional oxygen reservoir bag. The reservoir bag allows oxygen to collect while the patient is exhaling so that the oxygen concentration in the patient's next inhalation is increased. Depending on the oxygen source used to fill the reservoir bag, and also depending on the patient's settings, minute ventilation and inspiratory-expiratory ratio, concentrations of oxygen $> 80\%$ reportedly may be attained. Another new ventilator allows the clinician to turn off the bias flow, which also serves to allow a higher F_{IO_2} and to conserve oxygen. Many home ventilators can also now provide up to 60 cm H_2O of pressure support and 20 cm H_2O of PEEP. In actual practice, however, the maximum pressure support used at home is usually considerably less than 30 cm. Regardless of the ventilator technology, a determination must be made by the physician, respiratory therapist, and care-team to ensure that the patient is stable enough to be safely maintained at home.

Ventilator Design Issues for Home Mechanical Ventilation

There is considerable confusion on what actually constitutes a "ventilator." In its broadest sense, a positive pressure ventilator mimics spontaneous breathing by supplying pressure/volume to a patient during an inspiratory phase and releasing this pressure/volume during an expiratory phase. Other important features are a rate/inspiratory time setting and the ability to supply a level of expiratory pressure (PEEP). The FDA has classified these devices based on intended use and location of use (Table 6). Two of these codes are relatively new home ventilator codes, NOU and ONZ. NOU is the code for life support ventilators used at home. This code was created to

Table 4. Indications for Home Mechanical Ventilation

Indications for Home Noninvasive Ventilation
Individual has chronic stable or slowly progressive respiratory failure, as evidenced by:
Daytime CO_2 retention $\geq$ 50 mm Hg, with appropriately compensated pH, or
Mild daytime or nocturnal CO_2 retention (45–50 mm Hg) with symptoms attributable to hypoventilation (eg, morning headaches, restless sleep, nightmares, enuresis, daytime hypersomnolence)
Nocturnal hypoventilation or oxygen desaturation
And the following conditions have been met:
Individual has had optimal medical therapy for underlying respiratory disorders
Individual is able to protect airway and clear secretions adequately
Individual's reversible contributing factors have been treated (eg, obstructive sleep apnea, hypothyroidism, congestive heart failure, electrolyte disturbance)
The diagnosis is appropriate and may include:
Neuromuscular disease
Chest wall deformity
Central hypoventilation or obesity hypoventilation
Obstructive sleep apnea, and a failure to improve with nasal CPAP
COPD with severe hypercapnia or nocturnal desaturation
Indications for Home Invasive Ventilation
Individual meets indications for noninvasive ventilation *and* has:
Uncontrollable airway secretions despite use of noninvasive expiratory aids, or
Impaired swallowing leading to chronic aspiration and repeated pneumonias
Persistent symptomatic respiratory insufficiency and fails to tolerate or improve with noninvasive ventilation
Facial dysmorphisms
Patients placed on invasive ventilation during an emergency and who later refuse to change the system of ventilation or cannot be weaned
Individual needs > 20 h ventilatory support per day because of severely weakened or paralyzed respiratory muscles (eg, quadriplegia due to high spinal cord lesions or end-stage neuromuscular disease) and patient or provider prefers invasive ventilation

(Adapted from References 3 and 5.)

Table 5. Criteria for Stability and Discharge Readiness

Control of dyspnea
Airway stable
Can clear secretions and protect the airway
Acceptable arterial blood gases on $F_{IO_2} < 0.40$, and maintainable in the home
Stable metabolic and acid-base values
No acute infectious process
Medical regimen stable
No life-threatening cardiac dysfunction or arrhythmia
Other organ systems also stable
Nutrition adequate
In children, adequate growth and development
Can cope with the patient's physical and emotional needs at home
No predicted need for unscheduled or acute readmission or physician/clinician visit for 1 month
Contraindications for Home Ventilation
Medical condition potentially unstable
$F_{IO_2} > 0.40$ (relative)
PEEP > 10 cm H_2O
Continuous invasive monitoring needed
Tracheostoma not mature
Ventilator settings unstable
Airway resistance or compliance fluctuating
Patient does not want home ventilation
Physical environment unsafe
Health or safety hazards such as unsanitary conditions or fire danger
Utilities (eg, heat, air conditioning, electricity) inadequate (eg, amperage and grounded outlets)
Home care resources inadequate
Finances
Personnel
Medical follow-up
Patient unable to care for self when caregiver unavailable
Caregiver respite care inadequate
Number of competent caregivers inadequate ($\geq$ 2 caregivers required)

(Adapted from References 10 and 11.)

stipulate that this group of mechanical ventilators must be "tracked" by the durable medical equipment company so that, in the event of a recall, the device can be retrieved from the patient's home. ONZ is the latest home ventilator code. The ONZ device is a new type of volume assist ventilator that delivers a mix of 50 psi oxygen and room air, maintaining a minimum F_{IO_2} of approximately 36%. The device is intended to aid adult patients who are spontaneously breathing but who need some volume augmentation—especially during ambulation or exercise. The ONZ device can be used invasively or noninvasively.

An important engineering characteristic is whether an exhalation valve is required (patient-ventilator circuit is a closed system) or whether the system can operate in an open fashion with "leak compensation." The former design usually is associated with a higher level of support, tighter control of ventilator variables, and more extensive monitoring; the latter design is usually associated with the provision of only partial support (often with mask interfaces) and typically has fewer monitoring capabilities. These different features have led the reimbursement community to label the former a "ventilator" and the latter a "bi-level device" or, more commonly, a "respiratory assist device (RAD)" (Table 7).

The distinction between ventilator and RAD is becoming blurred. Newer ICU ventilator designs have ef-

Table 6. Ventilator Descriptions

FDA Product Code	FDA Description	Type of exhaust	Comments
CBK	Ventilator, continuous facility use	Exhalation valve	Commonly referred to as a "full" or "life" support device. Can manage patients who are ventilator dependent and/or not spontaneously breathing.
NOU	Continuous, ventilator, home use	Exhalation valve	Commonly referred to as a "full" or "life" support device. Can manage patients who are ventilator dependent and/or not spontaneously breathing. Must be locatable in the event of a recall.
MNT	Ventilator, continuous, minimal ventilatory support, facility use	Passive exhaust port	For facility use. Provides minimal ventilatory support; typically used to treat patients who require only some of the functions expected in critical care ventilators classified under CBK. Intended use statement specifies non-dependent and/or spontaneously breathing patients.
MNS	Ventilator, continuous, non-life-supporting	Passive exhaust port	Indicated for adults with obstructive sleep apnea and those who need ventilatory support during chronic or acute respiratory insufficiency. Patients should be expected to have no more than minor and transient adverse effects if ventilation/CPAP cannot be provided during extensive periods of time (eg, overnight).
BZD	Ventilator, non-continuous	Passive exhaust port	Indicated only for the treatment of adult obstructive sleep apnea.
ONZ	Ventilator, continuous	Passive exhaust port	Indicated for adults with respiratory insufficiency capable of spontaneously breathing a minimum of 3.5 mL/kg of predicted body weight.

Table 7. Ventilator and Respiratory Assist Device Medicare Coding Information

	Type of Rental	Code	Route	Description
Ventilator	Ongoing rental	E0450	Invasive	Volume control ventilator without pressure support, may include pressure control
	Ongoing rental	E0461	Noninvasive	Volume control ventilator without pressure support, may include pressure control
	Ongoing rental	E0463	Invasive	Pressure support ventilator with volume control, may include pressure control
	Ongoing rental	E0464	Noninvasive	Pressure support ventilator with volume control, may include pressure control
Respiratory assist device (RAD)	Capped rental	E0470	Noninvasive	RAD, bi-level pressure capability
	Capped rental	E0471	Noninvasive	RAD, bi-level pressure capability, with a backup rate
	Capped rental	E0472	Invasive	RAD, bi-level pressure capability, with a backup rate

Note that at the time of writing there is not yet a Centers for Medicare and Medicaid Services billing code for the ONZ ventilator.

fective leak compensation mechanisms that allow them to be used for partial support with a mask. Conversely, some newer RADs are designed to provide substantial support through a tracheostomy. Confusion develops when reimbursement schemes are tied to these device features rather than clinical needs and goals—a situation common in the payment policies of many payers, as described below.

With the advent of this new technology, clinicians will need to evaluate whether patients with a tracheostomy require a life-support ventilator or whether an approved RAD device can be appropriately used.[12] Certainly, there are some patients with tracheostomy who simply use a tracheostomy collar without the need for any ventilator support. Therefore it may be reasonable to view ventilator support for patients with tracheostomy on a continuum—from no support to partial support to full life-support technology, depending on factors such as the patient's condition, need for alarms, need for battery backup, and care environment. Another potential application for RADs that are approved for use with a tracheostomy may include patients who wish to enter a hospice pro-

gram—since hospice programs generally prohibit the use of life-support ventilators but traditionally allow the use of RADs.

Reimbursement Considerations for Home Ventilators and Related Supplies

Medicare part B covers medically necessary durable medical equipment, including ventilators and RADs. As noted above, the Medicare program has categorized devices with exhalation valves, adjustable ventilator rates, and alarms as mechanical ventilators, and places them in the group of devices requiring frequent and substantial service. Medicare will rent a mechanical ventilator for qualified patients as long as medical necessity exists. Conversely, Medicare places RADs in the capped rental category. For capped rental equipment, the durable medical equipment provider receives a monthly rental fee for 13 months, after which the ownership of the device can be transferred to the Medicare beneficiary. This is an important distinction—since patients on RADs typically do not receive home follow-up visits from a respiratory therapist, while patients on a mechanical ventilator are usually visited frequently for the first few months post-discharge, and then at least every 30–90 days thereafter—depending on patient need.

In addition to the device itself and the need for appropriate clinical support, patients on home mechanical ventilation also need supplies, power backup, and other durable medical equipment (eg, suction machine, nebulizer, home oxygen). Often a backup ventilator is clinically appropriate, though claims for a backup ventilator are frequently denied.[13] Working with payers and suppliers is an ongoing challenge for home caregivers. Details on managing these challenges effectively are given in the online supplement (see the supplementary materials at http://www.rcjournal.com).

How Many Ventilator Assisted Patients Are at Home in the United States?

An accurate count of the number of patients receiving home mechanical ventilation in the United States is unknown. In Europe, central databases of patients receiving home mechanical ventilation exist. An example is the EuroVent survey, a detailed survey in 16 European countries that identified patterns of home ventilator use.[14] For purposes of the survey, home mechanical ventilation was defined as invasive or noninvasive (regardless of whether a backup rate was provided). The use of ventilator adjuncts such as rocking beds was included. Patients with obstructive sleep apnea alone, or patients with a tracheostomy who did not use a ventilator, were not included. This survey found that home ventilator prevalence ranged from 0.1 to 17 per 100,000 people, with an average of 6.6 (Table 8).

Table 8. EuroVent Survey

Country	Estimated Ventilator Users	Estimated Prevalence (per 100,000)
Austria	300	3.8
Belgium	500	5.0
Denmark	500	9.6
Finland	450	8.7
France	10,000	17
Germany	5,000	6.5
Greece	70	0.6
Ireland	155	3.4
Italy	2,200	3.9
Netherlands	900	5.6
Norway	350	7.8
Poland	40	0.1
Portugal	933	9.3
Spain	2,500	6.3
Sweden	900	10
United Kingdom	2,320	4.1
All countries	27,118	6.6

(Data from Reference 14.)

Extrapolating this prevalence to the United States population in 2010 would suggest that the total American home ventilator population could be estimated to be 20,377 individuals.[15]

There are other ways to estimate home ventilator prevalence in the United States. Medicare claims data from 2010 indicates approximately 3,172 patients on home mechanical ventilation using an invasive interface, and 899 patients on home mechanical ventilation with a noninvasive interface. There are approximately 36,117 patients using a RAD without a backup rate (E0470), and 7,793 patients using a RAD with a backup rate (E0471). Since there is a code for the invasive use of RAD (E0472), it is assumed that the vast majority of patients on E0470 and E0471 devices are using the devices noninvasively. Unfortunately, there is no way to distinguish the number of E0470 patients who need ventilator support from those obstructive sleep apnea patients who were non-adherent to CPAP and were therefore moved to a bi-level device for comfort and enhanced adherence purposes. Taking these numbers together, it appears that there are roughly 47,981 Medicare patients receiving some form of home ventilation.[16] Of the total Medicare population using some form of RAD or ventilator, approximately 6.6% are using invasive ventilation (Table 9).

A 2006 report from St Elizabeth's Medical Center in Boston identified 464 individuals in their health system using prolonged mechanical ventilation, with 221 people, or 48%, receiving ventilation at home. Of the at-home group, 76% used a tracheostomy, while 14% used a mask.[17]

Table 9. Medicare Ventilator Support Claims Data 2010*

Code	Route	Description	Number of Patients
E0450	Invasive	Volume control ventilator without pressure support, may include pressure control	1,448
E0460	Noninvasive	Negative pressure ventilator (eg, chest cuirass, Porta-Lung)	22
E0461	Noninvasive	Volume control ventilator without pressure support, may include pressure control	182
E0463	Invasive	Pressure support ventilator with volume control may include pressure control	1,724
E0464	Noninvasive	Pressure support ventilator with volume control, may include pressure control	695
E0470	Noninvasive	Respiratory assist device (RAD), bi-level pressure capability	36,117
E0471	Noninvasive	RAD, bi-level pressure capability, with a backup rate	7,793
E0472	Invasive	RAD, bi-level pressure capability, with a backup rate	Data not available
Total all forms ventilation (RAD and all forms of mechanical ventilator)			47,981
Total invasive mechanical ventilator			3,172
Total noninvasive mechanical ventilator			899
Total noninvasive RAD			43,910
Invasive Ventilator Support by Region (%)			
United States (total all invasive/total all forms)			6.6
Europe (EuroVent survey data)†			13

* Centers for Medicare and Medicaid Services claims data, Noridian Administrative Services, accessed 7/29/2011.
† Data from Reference 14.

Table 10. Reported Home Mechanical Ventilator Failure

	No. (%)
Causes of Home Ventilator Failure Reports	
Defective equipment or mechanical failure	73 (39)
Improper care, damage, or tampering by caregivers	25 (13)
Functional equipment improperly used by caregivers	56 (30)
Functional equipment with change in patient's condition mimicking ventilator failure	5 (3)
No problem identified	30 (16)
Responses to Home Ventilator Failure Reports	
Ventilator replacement	84 (44)
Repair of defective part	11 (6)
Replacement of functioning ventilator solely for psychological comfort of the patient	27 (14)
Ventilator adjustments	40 (21)
Caregiver reeducated	13 (7)
Caregiver anxiety or distress reduced	5 (3)
No action required following assessment	7 (4)
Hospitalization	2 (1)

(Data from Reference 20.)

Table 11. Emergency Visits Over a 6-Month Period

Reason	Home Visits, No.
Ventilator not working	52
Technical issue (alarming, not reaching pressure, noisy)	43
Equipment required (tubing, filters, mask spares)	39
No fault	25
Circuit fitted incorrectly	9
Hospital requested exchange	9
Patient changed settings by mistake	8
Patient did not like replacement ventilator	2
Set up of ventilator at home	1
Total visits	188

(Data from Reference 21.)

According to the St Elizabeth report, the prevalence of long-term mechanical ventilation increased from 2.8 to 7.4 per 100,000 people over a 20 year period. Extrapolating these data to the United States population in 2010[15] suggests that the at-home mechanical ventilator population could then be estimated at 10,966 people, with 76%, or 8,334 people, using invasive ventilation.

A 2005 census of Massachusetts identified 197 pediatric patients who required prolonged mechanical ventilation. There were 137 of these patients (69%) who lived at home for the majority of the preceding 12 months. The study identified that 49% of the patients used invasive ventilation—so we can extrapolate that there were 67 invasively ventilated pediatric patients at home in 2005.[18] The pediatric population of Massachusetts, defined as people $\leq$ 18 years old, in 2009 (the closest year data were available) accounted for 21.7% of the total state population—or an estimated 1,420,835 children. This yields an estimated prevalence of 4.7 children per 100,000 children. Extrapolating these numbers to the 2010 total United States population would suggest the number of invasively ventilated children at home is 3,526.[15]

Finally, the state of Pennsylvania reported approximately 225 children on home ventilation in 2006, with about 80% (180 children) ventilated invasively.[6] Similar to the Massachusetts data above, this yields a prevalence of 6.4 children per 100,000 receiving home ventilation. Extrapolating this to the 2010 United States population[15] yields an estimated total prevalence of 4,802 children nationwide on invasive ventilation.

Table 12. Data on 8 Children Who Died

No.	Diagnosis	Years on Ventilator	Cause of Death	Years at Home
1	Spinal cord injury C2 quadriplegia	16 y	Ventilator disconnect, died at home	15 y, 7 mo
2	Spinal cord injury C2 quadriplegia	5 y	Cause unknown, found dead in bed at home	3 y, 8 mo
3	Spinal cord injury C2 quadriplegia	12 y, 10 mo	Bowel obstruction, peritonitis, died in hospital	6 y, 8 mo
4	Spinal cord injury quadriplegia	8 y, 6 mo	Unknown, died sitting in wheelchair after eating	0 y, 5 mo
5	Supplementary motor area seizures	5 y, 1 mo	Seizures, metabolic, died in hospital	4 y, 8 mo
6	Demyelinating neuropathy	4 y	Fall-accident in wheelchair, died while living at home	3 y, 1 mo
7	Myotubular myopathy	5 y	Overwhelming viral illness, died in hospital	3 y, 0 mo
8	Unknown myopathy	11 y, 10 mo	Ventilator disconnection, died at home	10 y, 5 mo

(Data from Reference 22.)

Table 13. Causes of Death in 47 Home Mechanically Ventilated Pediatric Patients

Cause of Death	No. (%)
Progression of reason for chronic respiratory failure or other underlying condition	16 (34)
Cardiac	10 (21)
Acute respiratory failure	4 (8.5)
Brain death	4 (8.5)
Infectious/sepsis/multiple organ dysfunction syndrome	4 (8.5)
Tracheal bleeding	4 (8.5)
Tracheal obstruction	4 (8.5)
Tracheostomy accident	1 (2)
Total	47 (100)

(Data from Reference 23.)

Table 14. Home Mechanical Ventilation Deaths Described in the FDA Manufacturer and User Facility Device Experience (MAUDE) Database—2010

Reported Cause of Death	No.
Patient passed away in his sleep. Mother says ventilator was functioning but did not alarm.	1
Alleged ventilator malfunction. No additional information.	1
Patient passed away while on ventilator. Ventilator was alarming and would not shut off until plugged in. Allegations of ventilator malfunction.	1
Patient passed away while on ventilator. Mother says no alarm. Sheriff's department investigating.	1
Patient had mucus plug while connected to ventilator, but the ventilator did not alarm.	1
Nurse discovered patient blue while connected to ventilator and began CPR. Ventilator alarmed.	1
Tracheostomy tube became dislodged. Ventilator allegedly did not alarm.	1
Tracheostomy tube became dislodged. Husband unsure if alarm going off or not, "as there were always alarms going off."	1
Caregiver claims patient passed away while on ventilator and alarms were delayed going off.	1
Patient expired while on ventilator. No allegation of ventilator malfunction. No further information.	1
Patient expired while on ventilator. Patient's family alleges ventilator did not alarm at the time of event.	1
Total	11

(Data from Reference 19.)

What Can Go Wrong During Home Mechanical Ventilation?

There are very limited data on home mechanical ventilator failure in the United States. A review of the FDA Manufacturer and User Facility Device Experience (MAUDE) database revealed over 150 alleged home mechanical ventilator malfunctions or failures in the year 2010.[19] A 1998 study of 150 ventilator-assisted patients ages 2–77 years reported 189 reported home ventilator failures over a 1 year period. The authors calculated this rate to be equivalent to one ventilator failure for every 1.25 years of continuous use. Table 10 details the reported causes of home mechanical ventilator failure, as well as the measures used to correct the suspected home ventilator failures. Interestingly, the author states that "equipment failure is not a frequent or serious problem for ventilator-assisted patients treated at home," although of the 69 patients receiving continuous ventilation, 74% had episodes of ventilator failure within the study year.[20]

A study from the United Kingdom evaluated the nature of calls to a support line for home ventilator patients. There were a total of 1,211 adult and pediatric patients: 1,199 on NIV and 12 on tracheostomy ventilation. Of the 1,211 patients, only 149 were 24 hour ventilator dependent. There was a mean of 528 daytime calls per month, and 14 nighttime calls per month, to the help line. Over a 6 month period, a total of 188 emergency home visits were made (Table 11).[21] Ventilator failure accounted for 28% of the visits to the home. This figure is especially problematic when considering that only approximately 12% of the study patients required full-time ventilation, and

therefore the number of home visits could reasonably be expected to increase substantially with a greater proportion of ventilator dependent patients.

Another study examined the cause of death for 8 patients who died while receiving home mechanical ventilation. It is worth noting that 2 of the 8 deaths were due to accidental disconnection of the ventilator circuit. One of the patients had been home on ventilation for 10 years, the other for 15 years, so presumably these families were well trained on proper ventilator management (Table 12).[22]

In southern California a retrospective observational cohort analysis of 228 children enrolled in a home mechanical ventilation program over 22 years (990 person years) was performed. Of the cohort, 47 of the 228 children died, and 41 were liberated from home mechanical ventilation. Neither age nor reasons for chronic respiratory failure were associated with shortened survival. Progression of underlying condition accounted for only 34% of the deaths; 49% of the deaths were unexpected. In the cohort, no deaths were caused by ventilator failure. There was evidence of notable risk from tracheostomy related events (Table 13).[23] The FDA MAUDE database described at least 11 patient deaths related to mechanical ventilation in the home in 2010.[19] Allegedly, in at least 5 of the 11 deaths, the ventilator did not alarm (Table 14).

A review of the FDA MAUDE database regarding home mechanical ventilation reveals that approximately 50 patients suffered a ventilator malfunction or failure that necessitated placing them on their backup ventilator, while at least 6 patients required cardiopulmonary resuscitation, during 2010.[19] The number of patients who had emergency need of their backup ventilator may be seriously underestimated, since the MAUDE report form does not ask specifically how the ventilator malfunction was addressed—so the person completing the form may or may not include detail about whether a backup ventilator was used.

It is unfortunate that there is no national registry for home ventilation patients in the United States,[24] making it impossible to know the exact number of patients requiring home ventilation. Although ventilator failures and serious incidents are to be reported to the FDA MAUDE database, without a central registry of ventilator patients it is impossible to determine a rate of ventilator failure/incidents for home ventilator patients. A central registry would allow for the development and monitoring of national home mechanical ventilator patient outcomes.

Summary

The preferred location for long-term mechanical ventilation is in the home, because costs are reduced, quality of life is enhanced, and integration into the community is maximized. The indications for both invasive and noninvasive mechanical ventilatory support in the home are increasing as technology and infrastructure support improves; however, reimbursement constraints make it challenging to provide home ventilator patients with the optimal equipment and services required. A central registry would allow for the development and monitoring of national home mechanical ventilator patient outcomes.

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Discussion

Carson: We were lamenting this morning how much of the care of the chronically critically ill patient is dictated by the payment structures. What can we do to support you? How do we get the key societies behind this?

King: Edwards and colleagues published a summary[1] of causes of death for pediatric patients on mechanical ventilation. They found only one death in the literature due to ventilator malfunction. I think that gives a false sense of security. The real question is how many times did the family absolutely need the backup ventilator due to the primary ventilator failing? And if the backup ventilator wasn't there, what would have happened?

I think we are probably underreporting to the FDA MAUDE [Manufacturer and User Facility Device Experience] database,[2] and we are underutilizing the data from MAUDE as well. It may not be clear to a lot of DME [durable medical equipment] companies that they are supposed to be submitting information to the MAUDE database, so who knows how many times families call and say "the ventilator broke" and the DME just brings them a new one, without completing the proper paperwork? We do know, from the database, that backup ventilators had to be used at least 72 times in 2010 due to the primary ventilator failing or malfunctioning! We know there were at least 150 reports of home ventilator failure or malfunction of some sort in 2010. But as to what the DMEs need? We need the physician groups to support, in writing, when backup ventilators are needed, so we can use that "ammunition" when dealing with the payers.

1. Edwards JD, Kun SS, Keens TG. Outcomes and causes of death in children on home mechanical ventilation via tracheostomy: an institutional and literature review. J Pediatr 2010;157(6):955-959.e2.
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MacIntyre: When do you use a RAD and when do you use a life-support ventilator, when they might have exactly the same settings?

King: That's why I'm bringing this up. This is a brand new issue that we could use some guidance on. In some countries, if the patient requires ventilator support more than X hours per day, he is placed on a life-support ventilator. There seems to be no consensus on the number of hours per day that constitutes "life-support ventilation." Some countries use 12 hours, some 14 hours, some more. To my knowledge, we don't have a specific rule. Ideally it should be based on the patient's condition and could be included on the certificate of medical necessity. This could be the determining factor for whether a life-support ventilator should be provided, and whether the patient needs a backup ventilator.

Another new area is the use of RADs to the tracheostomy. Previously we didn't have any RADs that were approved to be used with tracheostomies, so if a patient had a tracheostomy, it was a non-issue: you always put them on the life-support ventilator. Now we have a RAD that is FDA-approved for use to the tracheostomy, so we need guidance on which patients can safely use this technology, and when and if they need to transition to a life-support device.

MacIntyre: But, to be technical, the difference between a RAD and a ventilator is an exhalation valve, even though the RAD could do the same thing the ventilator does without the exhalation valve. So how do you decide? If I were in your shoes I'd say everybody needs an exhalation valve! What stops you from doing that?

King: There is now one life-support ventilator that can be used with what I call a "leak circuit," like a RAD. I don't think it's clear in the clinical community when to prescribe a RAD and when to prescribe a life-support ventilator for some patients with neuromuscular disease. At first, when the patient needs support only at night, certainly the RAD can be clinically appropriate and cost-effective. However, as the disease progresses and the patient begins using the RAD more and more hours per day, when should he transition to a life-support device?

In 2001, Lechtzin et al described an ALS [amyotrophic lateral sclerosis] patient who had been using NIV at home for over a year and died during a power failure. RAD patients often don't get much, if any, home follow-up to assess whether a transition to a life-support ventilator is needed. Life-support ventilator patients typically have batteries, priority power restoration, a backup ventilator, and frequent visits by a respiratory therapist.

1. Lechtzin N, Weiner CM, Clawson L. A fatal complication of noninvasive ventilation. N Engl J Med 2001;344(7):533.

MacIntyre: But you could set the device to be either one!

King: Exactly right. From a therapy perspective, I can set that particular life-support ventilator to deliver an IPAP and an EPAP and a backup rate, just like a RAD. From a therapy perspective, I can set that particular life-support ventilator to deliver an IPAP and an EPAP and a backup rate, using a leak circuit, just like a RAD. Or, I can set that device up with an exhalation valve to deliver traditional modes of ventilation. There is also a new RAD that includes an internal battery and that is approved for use with a tracheostomy. So we could use some physician guidance and clarification on when to use a RAD versus a life-support ventilator.

MacIntyre: I was trying to get a patient into an SNF [skilled nursing facility], a chronic patient with a tracheostomy on a ventilator receiving a modest level of pressure-support, and they said "we can't take him: he's on a conventional ventilator." Later I realized that they take patients on NIV at that SNF, so I called them back and said that we were going take him off the Puritan Bennett 840 and put him on a BPAP [bi-level positive airway pressure] machine attached to the tracheostomy, and they said, OK, we'll take him. It seemed so arbitrary, and the costs and reimbursement are so radically different. And SNF attitudes are radically different. This device that is approved for tracheostomies may be a tool to get people into SNFs much easier, because you can call it a RAD.

King: It's the same with hospice. Many hospice centers will accept a patient who is tracheostomized and using a RAD, but won't take that same patient on identical therapy with a life-support device. Again, we need physician guidance on this.

Hess: The problem is not unique to North Carolina. In the past, I've had patients with ALS who have been on 24/7 NIV who then elect to get a tracheostomy and they have to change their homecare company because the homecare company that was taking care of them on NIV doesn't take care of patients on ventilators. It's just continuing mechanical ventilation with a different interface.

King: That brings up an excellent point. Some of the companies that do RADs don't necessarily offer 24/7 on-call RT support, because a RAD is not considered a life-support device. It can be a case of "we'll be out in the morning or the next business day," versus with a life-support device, which has accreditation requirements for 24/7 respiratory support. That's an important distinction to check into as well.

Cheifetz: Angela, is it true that in the United States 50% of invasive home mechanical ventilation occurs in patients under 18 years of age? Do I correctly understand the data you presented?

King: To the best of my calculations, with very limited information, we have roughly 3,500 to 4,800 invasively ventilated pediatric patients. One study in Massachusetts indicated that 49% of the pediatric home patients were invasively ventilated.

Cheifetz: So it's about half. Thus, the percentage of pediatric home invasive ventilation is truly substantial. I realize that NIV may be a different story, but for invasive ventilation this percentage is much higher than I would have estimated.

King: The therapist from a good DME company is coming out about once a month to visit these pediatric patients. Unfortunately, the physician may give us a prescription to try "sprints" at home to try to get some of these kids off the ventilator—but due to the reimbursement system, it is mom and dad who have to supervise the sprints most of the time. It's such a shame that there is no reimbursement for the therapist to make home visits.

Cheifetz: The availability of pediatric-trained home health nurses and respiratory therapists is quite low. In my experience, the parents perform a substantial portion of the home care for these infants and children.

Carson: But I also think it's an explanation of why such a high percentage of the home ventilated patients are pediatric patients: because there are automatic caregivers, or at least one, whereas a 56-year-old may or may not have a capable caregiver.

Cheifetz: Yes, these pediatric patients do have automatic caregivers, but you have to remember that these caregivers are being taken out of the workforce. This can create substantial home stress, which I'll discuss tomorrow.

Bertuola:* I want to address the issue that in an SNF when you're trying to send a patient on a RAD, until recently we couldn't take a patient on a RAD when they're connected to a

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tracheostomy. So that has been an ongoing issue. Training becomes a huge issue, because the center might be just the traditional SNF, and now you're giving a patient a RAD and you have no external alarms. Is somebody going to hear it? Are they going to know how to address that alarm? We have a tendency to steer those patients to ventilators and put them on a ventilator, because of all those issues: you have to have skilled nursing staff who are trained in that type of care. There are a lot of challenges.

MacIntyre: I'm glad we have you here. Let me get this straight. You will take somebody on 15/5 of pressure support through a mask, but you would have fundamental difficulties with that same 15/5—same level of risk, same level of everything, but it's through a tracheostomy? The interface means a whole lot to your industry.

Bertuola: It does mean a lot. We look at those patients and ask are you venting them or are you treating OSA [obstructive sleep apnea]? If you're ventilating them, I steer them to a ventilator unit, because if that patient gets disconnected, we need to hear and answer that alarm. It's different than treating someone with OSA.

MacIntyre: I fully agree with you. The goal of therapy is critical: is it a life-support device that's going to cause harm if it's disconnected? Absolutely that's clear. But what about the patient who doesn't need it a lot, who can take it off to have dinner or watch TV, but does wear it at night? Why is the tracheostomy so important to a SNF? Why is it a critical barrier, given the patient with the same clinical needs?

Bertuola: Because, until recently, the FDA didn't allow the tracheostomy patient to be connected to a homecare BPAP in an SNF.

MacIntyre: But now you can't hide behind that anymore.

King: We used to define "life-support" as to the tracheostomy. If it was applied to the tracheostomy, it was considered life-support—but that definition doesn't really work anymore. Now we have 24 hour-a-day NIV patients, and we're fighting the insurance company, saying "Hey, he's life-support too: he needs a backup and RT visits." And there's the opposite end of the spectrum: we can now have a tracheostomy patient on a RAD. Is it life-support or not? We don't have a standard that we all agree on for defining "life-support."

Bertuola: I agree that we need a better definition, which would help define the amount of care that patient requires and the level of care we need to provide.

MacIntyre: Back home we define life-support versus non life-support in a very arbitrary way: we ask the physician who's in charge whether substantial harm would be likely if the patient became disconnected for a fairly brief period of time. If the answer is yes, then they're getting life support, and if the answer is no, then they're not. I realize it's vague, but at least in our institution that's the thought process we use in deciding whether to put somebody with NIV in an ICU or out on the floor.

Bertuola: Our challenge is that we're getting information from the hospital; we're not necessarily at the bedside and seeing what level of support is being provided to the patient, and whether they could tolerate being disconnected.