Risk Factors for Morbidity and Mortality in Pediatric Home Mechanical Ventilation

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Abstract

Background: Home mechanical ventilation (HMV) is increasingly used in children with chronic respiratory insufficiency, but data on incidence and type of adverse events are limited. *Setting:* Pediatric HMV program at a tertiary university hospital. *Methods:* The authors retrospectively analyzed the type and incidence of severe emergencies in a mixed pediatric HMV program. *Results:* In all, 295 patient-years of HMV in 54 patients could be analyzed. A total of 26 patients had neuromuscular disease. In 16 patients, mechanical ventilation was initiated at <1 year of age. A total of 45 children were ventilated via tracheostomy and 9 by nasal mask. This study identified 68 severe emergencies (0.2 per patient-year) leading to 4 deaths. Respiratory causes were found in 48 cases (including 15 tracheostomy-related and 3 ventilator failures). Only age, but not underlying diagnosis or mode of ventilation, correlated with incidence of emergencies. *Conclusions:* Pediatric HMV including all age and diagnostic groups shows a low incidence of emergencies.

Keywords

pediatric home mechanical ventilation, safety, morbidity, tracheostomy, noninvasive mechanical ventilation

Over the past 2 decades home mechanical ventilation (HMV) has become an established treatment in children with chronic respiratory insufficiency and is increasingly used in many countries.[1-6] Underlying etiologies in the majority of cases are neuromuscular diseases, including spinal muscular atrophy, muscular dystrophies, congenital myopathies, cervical spinal injury, and disorders of breathing control (eg, congenital central hypoventilation syndrome). Here, the underlying physiology of absent respiratory drive or muscular weakness leads to a restrictive type of respiratory compromise and allows mechanical ventilation with low pressures with no additional oxygen requirements and uncomplicated patient–ventilator synchrony. In this group of ventilator-dependent children, life expectancy may be limited not by respiratory insufficiency but by complications related to other organs (eg, myocardial in Duchenne patients).[7]

Other etiologies of chronic respiratory insufficiency and ventilator dependency in childhood are congenital syndromes with chest wall restriction, severe chronic lung disease of prematurity and chronic lung diseases refractory to medical treatment (eg, some cases of interstitial pneumonias, alveolar proteinosis, lung graft-versus-host disease, cystic fibrosis). In these diseases, mechanical ventilation is usually more challenging and prone to complications than in patients with neuromuscular diseases. High airway pressures, including elevated positive endexspiratory pressure and high oxygen, as well as patient–ventilator asynchrony, are encountered regularly. Increased morbidity and mortality resulting from complications of mechanical ventilation may reasonably be expected in this setting.

Nursing of technology-dependent children at home poses many challenges for family members and caregivers.[8] Common problems occurring in mechanical

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ventilation, for example, airway obstruction, hyperinflation, or hypoventilation have to be recognized and treated immediately. Most modern home mechanical ventilators and monitoring devices compare quite well with those used in the hospital and allow complex ventilation modes. These may be better adapted to individual patient needs but demand a thorough understanding by caregivers. In Germany, the majority of families caring for ventilator-dependent children are supplied with professional nurses covering at least the night hours. This may differ from the situation in other countries where pediatric HMV is performed. Lack of expert nursing care at home may lead to an increased incidence of life-threatening situations in pediatric HMV.

Noninvasive mechanical ventilation is increasingly used in pediatric HMV, and recent reports have demonstrated clinical efficiency and low mortality in neuromuscular patients.[5,6,9] Data on the safety profile of this ventilation mode compared with mechanical ventilation by tracheostomy are few and do not extend to diagnostic groups other than neuromuscular disease.[9-11] Furthermore, life-threatening emergencies occurring in HMV have been described,[10,12-19] but the incidence rate, including emergencies treated at home, is not known.

We therefore undertook a retrospective analysis of emergency situations occurring at home in children and adolescents with HMV by patient or caregiver questionnaire and hospital chart analysis. We speculated that patients with chest wall disorders or chronic lung diseases and patients with a low coverage of professional nurses may have an increased incidence of severe emergencies.

Methods

Description of the HMV Program

At our hospital, HMV care for ventilator-dependent children is initiated and led by pediatric intensivists in collaboration with pediatric pulmonologists and neurologists depending on the underlying etiology. After the decision to start HMV, caregivers are intensively instructed in life support measures, management of tracheostomies, including emergencies and recognition and management of ventilator problems. This regularly takes at least 4 weeks stay in hospital in tracheotomized and 2 weeks in noninvasively ventilated patients. During this stay, nursing staff for home care is organized. Nurses have to be trained in pediatric medicine and should be experienced in mechanical ventilation. This service is provided by a number of private nursing organizations and paid to a variable extent by standard health insurance.

Exclusively plastic cannulae are used in patients with tracheostomy. Interfaces for noninvasive mechanical ventilation are initially standard off the shelf followed by individually produced nasal masks. All patients are supplied with pulse oximetry and oxygen tank and ambu bag for emergency ventilation. Over the years, several brands of home mechanical ventilators have been used. Most patients are ventilated in a pressure-controlled mode.

After hospital discharge, telephone advice is offered 24 hours/days for 7 days/week, and check-up visits are fixed once a year (2 times a year in infants). In children with tracheostomy tubes, surveillance bronchoscopies are performed during these visits for early diagnosis of cannula-related problems. In addition, we perform electrocardiography and echocardiography to exclude right heart strain typically resulting from suboptimal ventilation as well as standard blood gas analysis in both awake and asleep states.

Study Population and Methods

Children and adolescents who were cared for in our HMV program up to May 2006 and were available for questioning were identified. We retrospectively asked for emergency situations from initiation of HMV up to the time point of the interview.

Patients were divided into diagnostic groups for the purpose of this study according to the following definitions: if muscular hypotonia or a disorder of ventilatory drive were the primary respiratory physiology patients were categorized as having neuromuscular disease; patients with airway stenoses or pulmonary disease as respiratory disease; all other as syndromatic diseases. These children had complex disorders, including skeletal syndromes with chest wall restriction, severe neurological compromise with spasticity, and recurrent aspirations.

Data were collected by questionnaire sent to parents or caretakers and hospital chart review. Telephone interviews were conducted when data were incomplete. The following data were obtained: age, gender, diagnosis, duration of mechanical ventilation, type of home care (nurses or family), and invasive versus noninvasive mechanical ventilation. Emergencies were defined as acute life-threatening deterioration of vital parameters requiring immediate intervention. Emergencies were subclassified according to need for acute medical service, need for resuscitation, and hospital admission. In 3 patients (a 3-year-old neuromuscular patient; a 5-year-old respiratory patient, and a 13-year-old patient with syndromatic disease), numerous emergencies handled at home were reported. For the purpose of statistical analysis this was counted as 4 emergencies as this was the upper limit of reported emergencies in the other patients.

Statistical analysis was performed by Poisson regression analysis. Parameters included age, diagnostic group,

Table 1. Patient Characteristics

Diagnosis	n	Age in Years (Mean/ Median)	Gender (Male/ Female)	Interface (Trach/ Mask)	Type of HMV (Continuous/ Intermittent)	Age at Start <1 Year	Time on HMV in Years (Mean/ Median)
Neuromuscular	26		16/10	20/6	15/11	5	5 mo-23 (7.4/5)
Spinal muscular atrophy	7	3-29 (16.6/18)	5/2	4/3	5/2	0	11 mo-23 (10.0/5)
Muscular dystrophy	3	9, 23, 27	2/1	3/0	3/0	0	5, 9, 13
Mitochondrial disease	3	3, 10, 16	1/2	3/0	2/1	2	2, 7, 15
CNS malformation	3	3, 12, 16	1/2	3/0	0/3	0	5 mo, 2, 10
Central congenital hypoventilation syndrome	3	8, 12, 16	3/0	3/0	1/ 2	2	4, 10, 15
HSMN	2	12, 15	1/1	0/2	0/2	0	2, 7
Spinal cord injury	2	3, 9	2/0	2/0	2/0	1	3, 5
Triose-phosphate isomerase deficiency	1	16	0/1	1/0	1/0	0	11
Multicore myopathy	1	11	1/0	0/1	0/1	0	5 mo
Tetraplegia of unknown cause	1	8	0/1	1/0	1/0	0	2
Respiratory	14	3-19 (9.1/8)	8/6	11/3	5/9	6	10 mo-14 (3.0/2.5)
BPD	4	3-9 (5.3/5.3)	2/2	4/0	3/1	2	10 mo, 11 mo, 4, 4
Airway stenosis	5	5-19 (11.2/9)	4/1	4/1	2/3	3	2-14
Restrictive[a] (lymphangiomatosis, S/P resuscitation, lung hypoplasia)	3	6, 12, 14	1/ 2	2/1	0/3	1	1, 2, 5
Pulmonary alveolar proteinosis	1	8	0/1	0/1	0/1	0	6
Cystic fibrosis + diaphragmatic hernia	1	8	1/0	1/0	0/1	0	38 mo
Syndromatic	14	2-26 (10.4/9)	9/5	14/0	6/8	5	3 mo-19 (6.3/6)
Arthrogryposis syndrome	5	2-13 (6.6/7)	2/3	5/0	3/2	4	1-12 (6/7)
Jeune syndrome	1	3	1/0	1/0	1/0	0	3 mo
Mucolipidosis/ mucopolysaccharidosis	2	4, 26	2/0	2/0	1/1	0	2, 8
Freeman–Sheldon syndrome	1	20	1/0	1/0	0/1	1	19
Infantile cerebral paresis	2	15, 17	1/1	2/0	1/1	0	5, 13
Unclassified syndromes[b]	3	7, 9, 11	2/1	3/0	0/3	0	1.5, 2, 7

Abbreviations: Trach, tracheostomy; HMV, home mechanical ventilation; mo, months; CNS, central nervous system; HSMN, hereditary sensorimotor neuropathy; BPD, bronchopulmonary dysplasia; S/P, status post.
[a]Respiratory restrictive: partial Di George syndrome plus epilepsy and bronchial stenoses; Lavy–Palmer–Merritt syndrome with lung hypoplasia and scoliosis; Gorham–Stout syndrome, S/P pulmonary radiation treatment, S/P resuscitation.
[b]Unclassified syndromes: laryngeal stenosis and unclassified cerebral and skeletal syndrome; severe CNS malformation plus endocrine abnormalities; multi-organ malformation syndrome.

interface, time on ventilator per day, and amount of nursing coverage.

Results

We could obtain complete data on 54 patients comprising 295 patient-years on home mechanical ventilation with a median age at survey of 10 years. In 16 patients, mechanical ventilation was initiated at <1 year of age. A total of 45 children were ventilated via tracheostomy and 9 by nasal mask. A change from nasal mask to tracheostomy was performed in 4 patients because of progression of their disease, whereas 4 patients could be weaned from mechanical ventilation. Further patient characteristics are listed in Table 1. The majority of patients were cared for by professional nurses at home, only 1 patient lived in a long-term facility. In total, 12 patients were cared for by nurses 24 × 7 hours, 11 patients

Table 2. Emergency Situations: Characteristics

	All (n)	Neuromuscular (Trach/Mask)	Respiratory (Trach/Mask)	Syndromatic (Trach/Mask)	Handled by Family or Nurse	Medical Rescue Team Alarmed	Resuscitation	Hospital Admission	Deaths
No. of patients with emergencies	30	13/2	5/0	10/0					
All emergencies	68	35/2	8/0	23/0	53	15		26	
Tracheostomy related	15	6	3	6	12	3	1	2	1 (tracheal bleeding)
Ventilator failures	3	3	0	0	3	0		0	
Other respiratory (infection, secretions, atelectasis)	30	17/2	5/0	6/0	21	9	3	16	1 (inactivated alarms)
Cardiac	1	1/0	0/0	0/0	0	1	1	1	1 (cardiac failure)
Neurological (seizures)	9	4/0	0/0	5/0	9	0		3	1
Others	10	4/0[a]	0/0	6/0[b]	8	2		4	
Call for medical rescue team	15	6/1	2/0	6/0					
Hospital admission	26	10/1	5/0	10/0					
Resuscitation	5	1/1	0/0	3/0				4	

Abbreviation: Trach, tracheal cannula.
[a]Electrolyte disturbance, 1; anaphylaxis, 1; unclear, 2.
[b]Gastrointestinal bleeding, 1; unclassified infection, 3; unclear, 2.

received no professional nursing care, and 30 patients had part-time home care.

A total of 26 patients had neuromuscular disease whereas 14 each had respiratory and syndromatic disease. Overall, 24 of 45 tracheotomized patients, compared with 1 of 9 noninvasively ventilated patients, were ventilated 24 hours per day. Of these 25 patients with continuous mechanical ventilation, 14 had neuromuscular disease. A total of 21 patients had daily mechanical ventilation from 4 to 12 hours.

Emergency situations were reported in 30 patients with more than one emergency occurring in 22 patients (see Table 2 for details). Therefore at least one emergency occurred per 4.6 patient-years.

In all, 15 of 26 (58%) children with neuromuscular disease suffered emergency situations compared with 5 of 14 (36%) with respiratory disease and 10 of 14 (71%) with syndromatic disease. However, there was no statistically significant difference between diagnostic subgroups in the incidence of emergency situations when appropriately analyzed as incidence per time of mechanical ventilation from initiation of ventilation to time point of analysis.

A total of 16 families reported failures of the ventilator resulting in 3 acute emergencies. None led to mortality or persistent adverse effects.

Overall, 15 tracheostomy-related severe emergencies were reported, one of them leading to death due to a massive tracheal arrosion bleeding in a patient with syndrome-associated tracheal scoliosis. This patient was under palliative care and arrived alive at the hospital, but the parents declined a surgical procedure.

There were 3 further deaths in our cohort. One patient died of neurological complications due to her underlying disease. One tracheostomized patient was found dead with disconnected ventilation tubing. Ventilator disconnection alarms did not function because of partial obstruction of the tubing (because the patient was lying on it). Pulse oximetry had been turned off because after years of false alarms, the family had decided to use it only during infections. The third patient died of cardiac failure due to mucopolysaccharidosis.

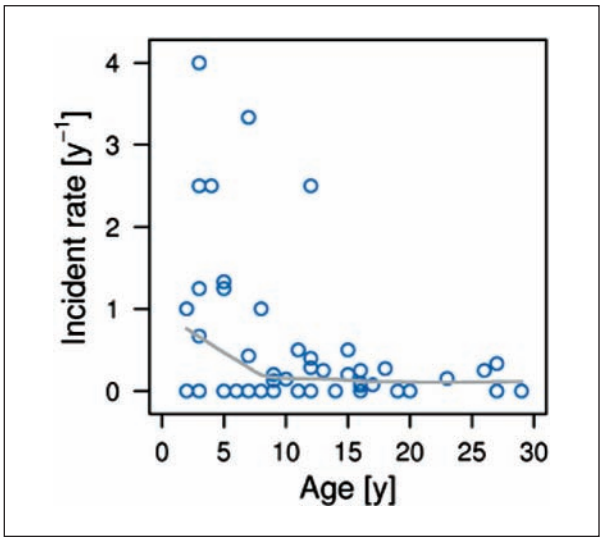

Figure 1. Incidence rate of severe emergencies (number of emergencies per patient-year of home mechanical ventilation) versus patient age ($P < .002$)
Line depicts locally weighted polynomial regression (loess) curve.

Severe respiratory problems, excluding tracheostomy-related and technology-related complications, were reported in 19 neuromuscular, 5 respiratory, and 6 syndromatic patients and were related to respiratory infections, increased secretions, and atelectasis. Neurological complications (mainly seizures) were noted in 9 and primary cardiac failure in 1 patient.

In only 15 cases, the emergency was severe enough that an acute medical care rescue team, including a physician, was called. This concerned 6 patients with neuromuscular disease, 2 patients with respiratory disease, and 6 patients with syndromatic disease, all with tracheostomy, and 1 patient with neuromuscular disease ventilated noninvasively. Five of these episodes involved resuscitation (2 neuromuscular patients, including 1 with noninvasive ventilation and 3 syndromatic patients) and one was terminated on request of the parents (acute cardiac failure in a mucopolysaccharidosis patient). A total of 26 episodes led to emergency hospital admission, whereas all the others could be managed at home without apparent sequelae.

There was a statistically significant indirect correlation of incidence rate of emergencies with age ($P < .002$ for multiple regression, see Figure 1). All other analyses did not show significance in a multivariate analysis (incidence rate vs type of ventilation, nasal mask vs tracheostomy and diagnostic group). When the incidence rate of emergencies was analyzed with respect to nursing coverage, no significant difference could be identified comparing patients with no care, <100 hours per week, ≥100 hours per week, and 24 hours for 7 days a week. This extended to all diagnostic subgroups.

Discussion

In this report, we demonstrate a low incidence of severe complications of pediatric HMV independent of underlying diagnosis, mode of ventilation interface, and nursing coverage. The study population comprises 295 patient-years and includes a substantial proportion of patients with complex respiratory physiology, which differs from earlier reports. Our analysis yielded an overall incidence of 68 life-threatening emergencies/295 patient years (eg, 0.27 per patient-year) defined by need for acute medical services, acute hospital admission, or resuscitation at home. An appreciable number of emergencies were handled at home without involvement of acute medical services.

Pediatric home mechanical ventilation is a therapeutic modality with complex medical, technological, social, and ethical issues[11,20-22] and a high level of financial and manpower resource use is typical. On the other hand, data on the achievements by this therapeutic modality concerning mortality and treatment-related morbidity are still scant.

Technology-dependent children have a high risk of hospital admission, including intensive care.[16] Available data on the hospital admission rate in pediatric HMV have shown rates ranging from lower than 1 to 1.53 per patient-year.[9,10,19] This included nonemergency admissions as well, whereas in a smaller series, the incidence of severe emergencies defined by death or hypoxic encephalopathy was 0.08 per patient-years.[18] Our data more specifically analyzed hospital admissions exclusively due to emergencies as well as those managed at home. Therefore, a direct comparison with earlier reports is limited by different inclusion criteria. Moreover, our population included a significant proportion of difficult-to-ventilate patients (eg, patients with lung disease, airway stenosis. or multiple organ pathology) whereas other series were composed of almost exclusively patients with neuromuscular disease or sleep apnea.

Death as outcome related to treatment complications was noted in only 2 cases, both of which could have been averted. A patient with tracheal arrosion bleeding due to the tracheal cannula arrived alive at the hospital and could have been saved by operation. This was declined by the parents as the patient was on palliative care because of his severe neurological disease. The second patient died because of failure of caretakers to implement pulse oximetry monitoring. We identified 5 cases of resuscitation at home leading to one further death due to underlying disease in a patient on palliative care

where resuscitation efforts were stopped on demand of the parents.

Earlier reports[10,12-14,15,17,18] have reported significantly higher mortality most probably linked to less optimal ventilator or monitoring technology compared with more modern series. For instance, in a series comprising 54 children reported in 1985,[10] there were 4 ventilator-associated deaths and in a report in 1992,[12] 3 deaths occurred in 32 children with congenital central hypoventilation syndrome due to cor pulmonale, which may be regarded as a sign of chronic underventilation.

The 64 emergencies resulted in only 31 (48%) emergency hospital admissions. The outcome of emergencies at home strongly depends on prompt detection and the competence and availability of caretakers. In our region, the majority of patients are provided with professional nursing care. Generally, nurses are trained in pediatric intensive care. This probably differs from the situation in several other countries with less resources where HMV is offered to children. In addition, every child is supplied with pulse oximetry and an oxygen tank for emergency bag and mask ventilation. Nevertheless, severe emergencies were not encountered more often in children without nursing care. Reasons may be better parental resources or competence or less severe respiratory insufficiency of the child, both of which may lead to less demand for nursing coverage. At our hospital, all parents are intensely trained in the management of complications from pediatric HMV over a couple of weeks during the initial hospital stay, which may be a major factor in reducing mortality of HMV independent of nursing coverage.

In a multivariate analysis, only age correlated with the incidence rate of emergencies. This confirms the general clinical impression that younger children are more difficult and technically demanding to ventilate because of higher physiological vulnerability and less respiratory reserve.[9] On the other hand, there could be a selection bias as more severely affected patients may die earlier and therefore are only included in the young age groups. We do not have evidence in favor of this as death rates were analyzed and were generally very low.

Surprisingly, we could not detect a significant effect of other possible risk factors on the incidence of emergencies (type of interface, continuous vs intermittent mechanical ventilation, diagnostic subgroup, and nursing coverage). Possibly patients with more severely compromised respiratory function or multiple problems were offered increased nursing coverage or more sophisticated technology and expanded monitoring. This could have prevented emergencies in the highest risk groups and confounded statistical analysis.

There are several well-known complications of tracheostomy, ranging from increased incidence of tracheobronchial infections to mechanical complications.[23,24] One of the most feared complications is tracheal arrosion with life-threatening bleeding. In our experience, this most often is related to the use of metal cannulas, which we therefore have abandoned. In our series, we encountered one case of tracheal arrosion bleeding in a patient using a plastic cannula. The patient had missed appointments and therefore the development of pressure lesions of the tracheal mucosa had not been identified. The bleeding could be stopped by placing the cuff of a tracheal cannula over the bleeding site but surgery was refused by the parents. In further 9 patients, cannula-related emergencies were mostly related to problems with cannula dislocation, which could be handled without the need of acute medical services and were without sequelae. This is in accordance with literature on home tracheostomy care in children[25] whereas specific data on tracheostomy-related problems in mechanically ventilated children at home are lacking.

There were 16 equipment/machine failures leading to 3 emergencies without any severe sequelae. This compares well with a report from a HMV program that included pediatric and adult patients where a failure rate of home ventilators amounting to 1 per 1.25 years of continuous use[26] was noted. Improper use or improper care of the ventilator was more often encountered than specific machine failure. Another report on ventilator malfunction confirmed a low incidence of severe complications.[27] In our patients, there were no complications, and all the emergencies were handled at home by caretakers. This may relate to the fact that all our ventilator-dependent children are provided with a second stand-by machine and oxygen tanks and ambu bags for manual ventilation.

Our study has several limitations. Its retrospective character over a long period of time (in some cases more than 20 years) allows substantial recall bias. Reporting bias may be introduced especially with emergencies handled at home, which in the majority of cases were reported by parents and only to a minor part by professional nurses. To capture emergencies as complete as possible, we applied a 2-tiered approach using caretaker questionnaire and hospital chart analysis. We used acute calls of medical services, resuscitation, and/or admission to hospital as surrogate parameters of severity of emergencies. We believe that it is improbable that we have missed relevant emergencies. Much less clear is how successful we were in identifying emergencies that were handled at home. These may nevertheless be life-threatening (eg, cannula dislocation) and may impose severe stress on parents and patients.

In conclusion, our data demonstrate a low incidence of severe complications in pediatric HMV. This extends to the most difficult to ventilate patients with restrictive and complex disease. Invasively and noninvasively ventilated patients demonstrated equal safety profiles. Only younger age correlated with an increased incidence of emergencies.

Declaration of Conflicting Interests

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