Outcomes and Causes of Death in Children on Home Mechanical Ventilation via Tracheostomy: An Institutional and Literature Review

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Objective To describe outcomes and causes of death in children on chronic positive-pressure ventilation via tracheostomy.

Study design We conducted a retrospective observational cohort of 228 children enrolled in an university-affiliated home mechanical ventilation (HMV) program over 22 years (990 person-years). Cumulative incidences of survival and liberation from HMV are presented. Time-to-events were compared by reason for chronic respiratory failure (CRF) and age and date of HMV initiation with Kaplan-Meier and Cox regression analyses. Circumstances of death are described.

Results Of our cohort, 47 of 228 children died, and 41 children were liberated from HMV. The 5-year cumulative incidences of survival and liberation were 80% and 24%, respectively. Being placed on HMV for chronic pulmonary disease was independently associated with liberation from HMV (hazard ratio, 7.38; 95% CI, 3.0-18.2; $P < .001$). Neither age nor reasons for CRF were associated with shortened survival. Progression of underlying condition accounted for only 34% of deaths; 49% of deaths were unexpected.

Conclusion Most children on HMV survive or were weaned off. However, a sizable number of children in our cohort died, and many deaths were unexpected and from causes not directly related to their primary reason for CRF. *(J Pediatr 2010;■:■-■).*

Home mechanical ventilation (HMV) via tracheostomy is perhaps the most demanding and risky of technology dependencies. Since the 1960s, HMV programs in the United States, and later across the globe, have demonstrated the relative safety and efficacy of HMV for supporting children with chronic respiratory failure outside of intensive care settings.[1] Even with this life-sustaining support, these children have complex chronic conditions, often with associated co-morbidities, that put them at risk for critical illness and death.

Nineteen earlier institutional surveys have reported on the outcomes of 621 pediatric patients on chronic PPV via tracheostomy in 6 countries, with follow-up ranging from 4.5 to 25 years (**Table I**; available at www.jpeds.com).[2-20] Incidences of mortality in these studies ranged from 0 to 43%; incidences of liberation from PPV were 0 to 52%. These surveys had relatively small cohorts (6-101 patients), and most studies reported little on the circumstances of deaths in their cohorts. Some commentaries have suggested that the survival of these children is primarily influenced by the clinical course of their underlying disease.[3,12,21] In contrast, our anecdotal experience suggested that the deaths in our patients were from more varied causes and often not anticipated.

Therefore, we conducted a retrospective cohort analysis of our relatively large population of children and young adults on chronic positive-pressure ventilation (PPV) via tracheostomy to describe their outcomes and the circumstances of their deaths.

Methods

We performed a retrospective chart review of all patients on HMV who received full- or part-time chronic PPV via tracheostomy and were observed at Children's Hospital Los Angeles (CHLA) between November 1977 and April 2009. The CHLA Institutional Review Board approved this review. Data was extracted from paper charts, the Pediatric Intensive Care Unit database (Microsoft Access, Microsoft Corporation, Redmond, Washington), and the hospital database (Knowledge, Information, Decision Support, Cerner Corporation, Kansas City, Missouri). Limited, additional patient-specific information was obtained

CHS	Central hypoventilation syndromes
CHLA	Children's Hospital Los Angeles
CI	Confidence interval
CPD	Chronic pulmonary diseases
CRF	Chronic respiratory failure
HR	Hazard ratio
HMV	Home mechanical ventilation
PPV	Positive-pressure ventilation
VMW	Ventilatory muscle weakness

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Table III. Survival and weaning status of 228 home mechanical ventilation patients by reason for chronic respiratory failure

			Weaned		Died	
Cause of CRF	Total patients n (%)	Alive on HMV n (%)	n (%)	95% CI	n (%)	95% CI
CPD	120 (52.6)	62 (51.7)	35 (29.1)	21.2-38.2	23 (19.2)	12.6-27.4
VMW	62 (27.2)	45 (72.6)	4 (6.4)	1.8-15.7	13 (21)	11.7-33.2
CHS	46 (20.2)	33 (71.7)	2 (4.3)	0.5-14.8	11 (23.9)	12.6-38.8
Total	228	140 (61.4)	41 (18)	13.2-23.6	47 (20.6)	15.6-26.5

from the HMV program coordinator (S.K.) and the CHLA pediatric pulmonology faculty.

Data was collected on each patient's primary cause of chronic respiratory failure (CRF) and indication for HMV, sex, age at initiation of HMV, date of initiation, survival status, and, when applicable, age at death or liberation from HMV. CRF was defined as full- or part-time ventilator dependence for at least 1 month after repeated, failed attempts to wean from assisted ventilation in a person without superimposed acute respiratory disease. Causes of CRF were classified in 3 subgroups: chronic pulmonary diseases (CPD), ventilatory muscle weakness (VMW), and central hypoventilation syndromes (CHS). Examples of diagnoses for each subgroup can be found in Table II (available at www.jpeds.com). Patients with multi-factorial reasons for CRF were discussed with the program coordinator and the faculty member who knew the patient the best to decide their primary cause of CRF.

Survival status was determined to be alive on HMV, weaned off HMV, or deceased. For deceased subjects, we performed a more thorough review of their medical records to collect additional data on cause of death, place of death, and whether the death was expected. Expected death was defined as an attending physician documenting that the child was at risk for death outside of the hospital or, for inpatients, that the patient was at risk for death or that death was anticipated within one week. Documentation of anticipated death was not considered to be expected when the death followed acute, severe deterioration within 24 hours.

Patients who were >18 years of age at the time of HMV initiation or with insufficient information were excluded. Children who died after being liberated from HMV were analyzed in the weaned off HMV group.

Data are presented as medians and interquartile ranges (IQR) or as proportions and 95% CIs. Univariate analysis of patient characteristics and causes of death for different reasons for CRF were tested with the Fisher exact test and Kruskal-Wallis rank test, as appropriate. Kaplan-Meier survival and cumulative incidence of liberation from HMV curves are presented and compared with the log-rank test. The relationships among reason for CRF, age at HMV initiation, year of initiation, survival, and liberation from HMV were analyzed with Cox proportional hazards models. Statistical significance was determined with a P value of .05 and by constructing 95% CIs. Statistical analyses and graphs were performed with Stata software version 11 (StataCorp LP, College Station, Texas).

Results

Since 1977, the CHLA HMV program has cared for 388 children and young adults receiving full- or part-time chronic PPV via tracheostomy. One hundred forty-two patients (36%) were excluded because of insufficient information or loss to follow-up. The cause of CRF and the dates of HMV initiation, liberation, or death could not be confirmed in the medical record for 126 children (32%). Of these children, 73 (19%) were initiated on HMV before July 1987—the earliest date a study patient was started on HMV. Sixteen patients (4%) were lost to follow-up after 1987. Eighteen additional patients (5%) were excluded because they were initiated on HMV after 18 years of age. The remaining 228 patients (59%) comprised our study population and contributed 990 cumulative person-years of follow-up. The longest duration of follow-up was 212 months. Ninety-eight of 228 children (43%) were female.

Seventeen of 228 patients (7.5%) had a multi-factorial etiology of CRF—14 (6%) had both CPD and CHS diagnoses, and 3 (1.5%) had CPD and VMW diagnoses. For 12 patients, the non-CPD diagnosis was determined to be the primary cause of CRF.

The median age of initiation of HMV was 11 months (IQR, 6-41.5 months; range 1-210 months). When stratified by reason for CRF, children with CPD were initiated on HMV at significantly younger ages (median, 8 months) than children with VMW or CHS (median, 21.5 and 25.5 months, respectively; Kruskal-Wallis rank test, 17.1; $P = .0002$).

Table III details the survival status of our cohort with the incidences of all-cause mortality and liberation from HMV, with their 95% CIs, by reason for CRF. One hundred forty patients (61%) remain alive on HMV. For the entire cohort, the 5- and 10-year cumulative incidences of survival were 80% (95% CI, 73%-85%) and 63% (95% CI, 51%-73%), respectively.

Forty-one patients (18%) were liberated from HMV. Excluding patients who died, the 5- and 10-year incidences of liberation were 24% (95% CI, 18%-32%) and 29% (95% CI, 21-39%), respectively. Thirty-seven of 41 liberated patients (90%) were weaned within 5 years of initiation. All liberated patients were weaned to tracheostomy collar, and none required non-invasive ventilatory support. Two patients with

congenital central hypoventilation syndrome are now dependent on diaphragm pacers. The median time to liberation from HMV was 28.5 months (IQR, 16.5-48 months). Figure 1 illustrates the cumulative incidence curves of HMV liberation by reason for CRF. There was a significant difference in time to liberation from HMV in these subgroups (log rank, 31.9; $P < .0001$). Cox proportional hazard modeling showed that CPD as the reason for CRF is independently associated with earlier weaning (hazard ratio [HR], 7.38; 95% CI, 3.0-18.2; $P < .001$), after controlling for other causes of CRF, age at initiation, and date of initiation. Age of initiation is not independently associated with earlier liberation with Cox modeling ($P = .12$).

Forty-seven of 228 patients (21%) died while on HMV. Their median time on HMV was 22 months (IQR, 8-60; range, 1-176 months). The incidence rate of death was 0.47 per 10 person-years (95% CI, 0.35-0.63). Figure 2 illustrates the Kaplan-Meier survival curves for the cohort by reason for CRF. The subgroups' survival curves are not significantly different (log rank, 2.77; $P = .25$). Cox regression demonstrated that earlier date of HMV initiation is independently associated with shortened survival (HR, 1.06; 95% CI, 1.004-1.13; $P = .036$). Neither reason for CRF nor age at initiation are independently associated with shortened survival ($P = .07$ and $P = .41$, respectively).

Table IV details the causes of death for these patients by reason for CRF. Three deaths caused by progression of underlying disease involved withdrawal of support; these deaths were evenly divided in the causes of CRF. Nine deaths (19%) were tracheostomy-related. No deaths were caused by ventilator malfunction. Twenty-four deaths (51%) were expected, per the documentation of attending physicians. No clinically determinable cause, beyond cardiac or brain death, was found in 5 of the unexpected deaths (11%), although autopsies were not performed or the results were inconclusive. Three patients (1.3%) who were liberated from HMV subsequently died 2 to 9 years later. Two of these patients died as a result of their underlying cardiac disease; the third patient died unexpectedly at home for unclear reasons.

Twenty-one patients (45%) died in an intensive care unit; 8 patients (17%) died on a ventilator ward; and 7 patients died (15%) in an emergency department. Eleven patients (23%) died outside of a hospital; 9 patients (19%) died at home and two patients (4%) at a chronic care facility.

Discussion

Our survey of 228 children and adolescents on HMV via tracheostomy represents the largest pediatric cohort reported to date and the first to apply proportional hazards analysis. The incidences of mortality and liberation from HMV are comparable with other studies with similar periods of follow-up (Table I). Our cohort had a smaller proportion of patients who died outside of a hospital—23% versus the composite 39% (15 of 39 dead patients) that is reported in 4 earlier papers.[4,10,14,20] Besides having a greater proportion of cardiac and tracheostomy-related deaths, the causes of death in our cohort are comparable with those in the 13 studies that reported the causes of their patients' deaths (Table V; available at www.jpeds.com).[2,3,5-7,9-12,14,16,17,19]

Thirty-five patients (15%) total and 12 dead patients (25%) had corrected or palliated congenital heart disease and are discussed elsewhere.[22] Ten of these patients died of cardiac-related reasons. In this analysis, the cause of death for 7 of these patients was designated as progression of underlying illness, 3 deaths were designated as cardiac (but not directly related to their original cardiac lesion), one death was from sepsis, and the last death was from tracheal bleeding. Of the remaining 7 patients who had cardiac-related deaths but no congenital heart disease, 3 died from

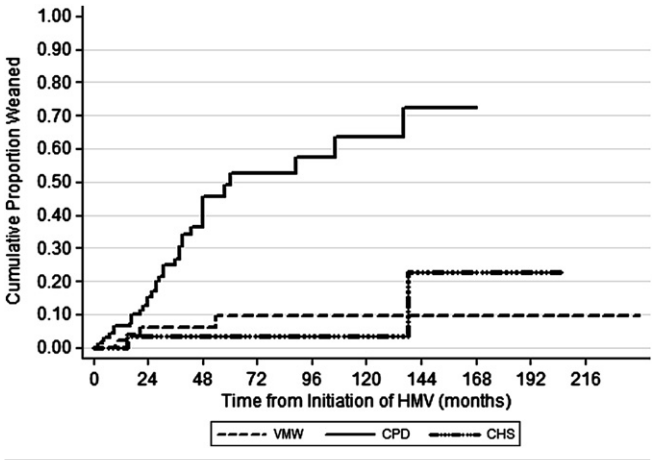

Figure 1. Cumulative incidence curves of liberation from HMV for 181 children by reason for CRF.

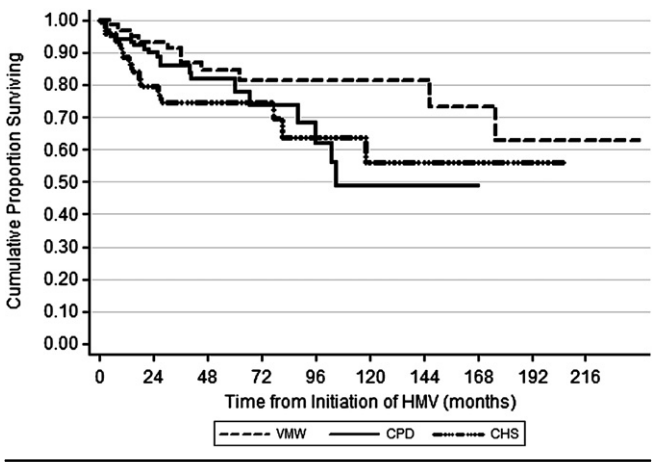

Figure 2. Kaplan-Meier survival curves for 228 children on HMV by reason for CRF.

Table IV. Causes of death in 47 home mechanical ventilation patients by reason for chronic respiratory failure

Cause of death	Total n (%)	CPD n (%)	VMW n (%)	CHS n (%)	*P* value*
Progression of reason for CRF or other underlying condition	16 (34)	12 (26)	1 (2)	3 (6)	.13
Cardiac	10 (21)	6 (13)	3 (6)	1 (2)	.81
Acute respiratory failure	4 (8.5)	1 (2)	2 (4)	1 (2)	.68
Brain death	4 (8.5)	1 (2)	1 (2)	2 (4)	.68
Infectious/sepsis/MODS	4 (8.5)	1 (2)	1 (2)	2 (4)	.68
Tracheal bleeding	4 (8.5)	2 (4)	1 (2)	1 (2)	1
Tracheal obstruction	4 (8.5)	–	4 (8.5)	–	.01
Tracheostomy accident	1 (2)	–	–	1 (2)	.25
Total	47	23 (49)	13 (28)	11 (23)	

MODS, Multiple organ dysfunction syndrome.
*Fisher exact test.

arrhythmias induced by electrolyte abnormalities. Four patients died unexpectedly, connected to their ventilators, with no evidence of airway obstruction or antecedent illness. Their causes of death were classified as cardiac-related by the pronouncing physicians or by autopsy.

In contrast to earlier commentaries that mortality in this population is largely related to their underlying diagnosis, our review of causes of death in the literature and our cohort suggest a more nuanced conclusion. Death by progression of underlying disease was the cause in only approximately 35% of reported deaths, both here and in the literature. The cause of death could not be fully explained in 10% to 15% of reported deaths. Also, approximately half the deaths in our cohort were unexpected. In 4 earlier studies that provide such detail, a composite 43% of their 35 deaths (incidences ranged from 25%-50%) were described as unexpected.[2, 9,12,16] Serwint and Nellis similarly found that deaths in children with chronic conditions were often unexpected.[23] Our experience suggests that many of the unexpected deaths, those caused by reasons other than progression of underlying condition, and, likely, the unexplainable deaths are related to the nonpulmonary co-morbidities these patients commonly have.

Most of our patients were weaned within 5 years from HMV initiation; only 4 patients (10%) were weaned after that point. In patients with central hypoventilation syndromes, we anticipate the number of liberated patients to grow with increasing use of new technologies, such as diaphragmatic pacing.[24] We did not review the incidence or timing of tracheostomy decannulation in our weaned cohort. Some patients continued to require supplemental oxygen via tracheostomy collar, although none required noninvasive or other ventilatory support except two patients with diaphragm pacers.

Regression analysis suggests that CPD patients have a greater likelihood of liberation from HMV (HR, 7.38) compared with patients in the other two CRF subgroups. We believe this is partly explainable by the large proportion of bronchopulmonary dysplasia patients in the CPD group. Earlier studies showed that patients with bronchopulmonary dysplasia are more likely than other diagnostic groups to wean from HMV.[12,25,26]

Earlier date of HMV initiation is a modest independent predictor of shortened survival (HR, 1.06), which may suggest that we became better at caring for these children and their complex conditions. Similar to earlier studies, we found no significant difference in survival time by diagnostic category.[2,11] Our univariate analysis of causes of death by reason for CRF did find that tracheal obstruction (ie, mucus plugging in all cases) was significantly different in the subgroups ($P = .01$). All these deaths occurred in VMW patients, possibly related to their ineffective cough.

The relative safety of HMV or, more specifically, the absence of home ventilator malfunction has been prospectively observed.[27] In our cohort, no deaths were caused by ventilator malfunction. Indeed, we found only one reported death from positive-pressure "ventilator failure" in the literature.[17] However, there does seem to be evidence of notable risk from preventable tracheostomy-related events. Eight percent of deaths reported in the literature and 19% in our cohort were tracheostomy-related. In our cohort, these avoidable causes of death included mucus plugging, tracheostomy being replaced into a false track, and hemorrhage of tracheal granulomas. To prepare families for the care of their child on HMV and prevent life-threatening accidents, we provide caregivers with extensive training and other supports before the child is discharged from the hospital (**Table VI**; available at www.jpeds.com).

Many patients had multiple co-morbidities that arguably contributed to their CRF. Seventeen patients had more than one possible reason for CRF. Within the limits of our retrospective review, we identified their primary reason for HMV. Our results, stratified by CRF, did not change significantly when statistical analyses were repeated with the alternative cause of CRF for these 17 patients.

There were some distinctive features of our outcome analysis and comparison with earlier studies. In general, children on HMV are a heterogeneous population with different indications for chronic ventilatory support and a variation in number, type, and severity of co-morbidities. Likewise, earlier studies encompassed a wide range of subgroups, follow-up periods, and geographic sites. Additionally, they represent different periods with different technology. These differences precluded a true meta-analysis of these studies. As with earlier studies, our own study is a retrospective survey of one institution's experience. Our analysis included only 77% of the pediatric patients on HMV that we have observed since July 1987. Although we do not believe the excluded patients are different from those in the study, this

certainly may have introduced selection bias. We did not directly compare children on HMV who died or were liberated from HMV to children who did not. Therefore, beyond etiology of chronic respiratory failure, age at initiation, and date of initiation, we cannot make inferences on factors that are associated with death or successful weaning.

Even with increasing use of chronic non-invasive ventilatory support, we anticipate that the use of chronic PPV via tracheostomy will become more prevalent in the future.[7,19,28] Although HMV can be a relatively safe and effective means of assisting respiratory function, it is neither curative nor does it prevent acute illness or the progression of underlying and co-morbid conditions. These limitations are reflected in the evidence that perhaps one-fifth of these patients die within 5 years of initiation and that a sizable proportion of these deaths are unexpected and likely caused by co-morbidities or critical illnesses that are not directly related to their reason to be on HMV.

Outcomes data are imperative for the families considering HMV for their child and for the healthcare professionals who help families make decisions and care for the child before and after HMV initiation. Multi-institutional outcomes research with more stratification of factors that likely mediate mortality and successful weaning is needed. Similarly, all decision-makers and caregivers need to understand and appreciate the limitations and risks of HMV. Only with robust data and realistic expectations can good decisions be made for children with chronic respiratory failure. ■

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References

1. Make BJ, Hill NS, Goldberg AI, Bach JR, Criner GJ, Dunne PE, et al. Mechanical ventilation beyond the intensive care unit. Report of a consensus conference of the American College of Chest Physicians. Chest 1998;113:289-344.
2. Frates RC, Jr., Splaingard ML, Smith EO, Harrison GM. Outcome of home mechanical ventilation in children. J Pediatr 1985;106:850-6.
3. Schreiner MS, Downes JJ, Kettrick RG, Ise C, Voit R. Chronic respiratory failure in infants with prolonged ventilator dependency. JAMA 1987;258:3398-404.
4. Oren J, Kelly DH, Shannon DC. Long-term follow-up of children with congenital central hypoventilation syndrome. Pediatrics 1987;80:375-80.
5. Iannaccone ST, Guilfoile T. Long-term mechanical ventilation in infants with neuromuscular disease. J Child Neurol 1988;3:30-2.
6. Gilgoff IS, Kahlstrom E, MacLaughlin E, Keens TG. Long-term ventilatory support in spinal muscular atrophy. J Pediatr 1989;115:904-9.
7. Robinson RO. Ventilator dependency in the United Kingdom. Arch Dis Child 1990;65:1235-6.
8. Fields AI, Coble DH, Pollack MM, Kaufman J. Outcome of home care for technology-dependent children: success of an independent, community-based case management model. Pediatr Pulmonol 1991;11:310-7.
9. Mallory GB, Jr., Stillwell PC. The ventilator-dependent child: issues in diagnosis and management. Arch Phys Med Rehabil 1991;72:43-55.
10. Marcus CL, Jansen MT, Poulsen MK, Keens SE, Nield TA, Lipsker LE, et al. Medical and psychosocial outcome of children with congenital central hypoventilation syndrome. J Pediatr 1991;119:888-95.
11. Canlas-Yamsuan M, Sanchez I, Kesselman M, Chernick V. Morbidity and mortality patterns of ventilator-dependent children in a home care program. Clin Pediatr (Phila) 1993;32:706-13.
12. Wheeler WB, Maguire EL, Kurachek SC, Lobas JG, Fugate JH, McNamara JJ. Chronic respiratory failure of infancy and childhood: clinical outcomes based on underlying etiology. Pediatr Pulmonol 1994;17:1-5.
13. Sasaki M, Sugai K, Fukumizu M, Hanaoka S, Kaga M. Mechanical ventilation in severe childhood neurological disorders. Brain Dev 2001;23:796-800.
14. Appierto L, Cori M, Bianchi R, Onofri A, Catena S, Ferrari M, et al. Home care for chronic respiratory failure in children: 15 years experience. Paediatr Anaesth 2002;12:345-50.
15. O'Brien JE, Dumas HM, Haley SM, O'Neil ME, Renn M, Bartolacci TE, et al. Clinical findings and resource use of infants and toddlers dependent on oxygen and ventilators. Clin Pediatr (Phila) 2002;41:155-62.
16. Gilgoff RL, Gilgoff IS. Long-term follow-up of home mechanical ventilation in young children with spinal cord injury and neuromuscular conditions. J Pediatr 2003;142:476-80.
17. Nelson VS, Dixon PJ, Warschausky SA. Long-term outcome of children with high tetraplegia and ventilator dependence. J Spinal Cord Med 2004;27(Suppl. 1):S93-7.
18. Edwards EA, O'Toole M, Wallis C. Sending children home on tracheostomy dependent ventilation: pitfalls and outcomes. Arch Dis Child 2004;89:251-5.
19. Gowans M, Keenan HT, Bratton SL. The population prevalence of children receiving invasive home ventilation in Utah. Pediatr Pulmonol 2007;42:231-6.
20. Oktem S, Ersu R, Uyan ZS, Cakir E, Karakoc F, Karadag B, et al. Home ventilation for children with chronic respiratory failure in Istanbul. Respiration 2008;76:76-81.
21. Amin RS, Fitton CM. Tracheostomy and home ventilation in children. Semin Neonatol 2003;8:127-35.
22. Edwards JD, Kun SS, Keens TG, Khemani RG, Moromisato DY. Children with corrected or palliated congenital heart disease on home mechanical ventilation. Pediatr Pulmonol 2010;45:645-9.
23. Serwint JR, Nellis ME. Deaths of pediatric patients: relevance to their medical home, an urban primary care clinic. Pediatrics 2005;115:57-63.
24. Chen ML, Tablizo MA, Kun S, Keens TG. Diaphragm pacers as a treatment for congenital central hypoventilation syndrome. Expert Rev Med Devices 2005;2:577-85.
25. Kharasch VS, Haley SM, Dumas HM, Ludlow LH, O'Brien JE. Oxygen and ventilator weaning during inpatient pediatric pulmonary rehabilitation. Pediatr Pulmonol 2003;35:280-7.
26. O'Brien JE, Dumas HM, Haley SM, Ladenheim B, Mast J, Burke SA, et al. Ventilator weaning outcomes in chronic respiratory failure in children. Int J Rehabil Res 2007;30:171-4.
27. Srinivasan S, Doty SM, White TR, Segura VH, Jansen MT, Davidson Ward SL, et al. Frequency, causes, and outcome of home ventilator failure. Chest 1998;114:1363-7.
28. Graham RJ, Fleegler EW, Robinson WM. Chronic ventilator need in the community: a 2005 pediatric census of Massachusetts. Pediatrics 2007;119:e1280-7.

Table I. Outcomes of pediatric patients on chronic positive-pressure ventilation via tracheostomy in 19 previously published studies

Article	n	Alive on HMV n (%)	Died on HMV n (%)	Weaned off HMV n (%)	Follow-up (years)	Comments
Frates et al[2] 1985	33	16 (48)	10 (30)	7 (22)	20	13 of 29 patients had spinal cord injuries
Oren et al[4] 1987	6	5 (83)	-	1 (17)	14	All patients with CCHS
Schreiner et al[3] 1987	101	18 (18)	20 (20)	63 (62)	18	Included 8 children on NPV; all initiated <1 year of age; 10 patients died after liberation from HMV; 3-year survival rate = 71%.
Iannaccone et al[5] 1988	7	3 (43)	3 (43)	1 (14)	4	All patients with neuromuscular disease; HMV initiated before age 2 years
Gilgoff et al[6] 1989	14	12 (86)	2 (14)	-	24	All patients with spinal muscle atrophy; 10 patients only required nighttime ventilation.
Robinson[7] 1990	35	27 (77)	6 (17)	2 (6)	14	United Kingdom
Fields et al[8] 1991	23	11 (48)	5 (22)	7 (30)	4.5	
Mallory et al[9] 1991	28	16 (57)	12 (43)	-	6	
Marcus et al[10] 1991	13	12 (92)	1 (8)	-	12	All patients with CCHS
Canlas-Yamsuan et al[11] 1993	19	9 (47)	6 (32)	4 (15)	13.5	Canada
Wheeler et al[12] 1994	55	32 (58)	4 (7)	19 (36)	5	
Sasaki et al[13] 2001	25	18 (72)	7 (28)	-	10.5	Japan; all had neurological disorders
Appierto et al[14] 2002	34	25 (74)	9 (26)	-	15	Italy
O'Brien et al[15] 2002	17	13 (76)	-	4 (24)	6.5	All in inpatient pediatric pulmonary rehabilitation program
Gilgoff et al[16] 2003	39	31 (79)	8 (21)	-	20	All patients with spinal cord injury or neuromuscular conditions.
Edwards et al[18] 2004	39	32 (82)	7 (18)	-	8	United Kingdom
Nelson et al[17] 2004	45	25 (55)	16 (36)	4 (9)	25	All patients with spinal cord injury; included 4 children on NPV or mask ventilation
Gowans et al[19] 2007	77	47 (61)	13 (17)	17 (22)	9	
Oktem et al[20] 2008	11	5 (46)	4 (36)	2 (18)	5	Turkey
Total	621	357	133	131		

When studies reported on pediatric and adult subjects or on other forms of chronic ventilatory support, information pertinent to patients initiated on chronic PPV before 18 years of age was extracted. Two studies[3,17] had a small (7.9%-9.5%) proportion of patients on chronic ventilatory support other than PPV whose outcome information could not be separated from the larger PPV cohort and were included.

CCHS, Congenital central hypoventilation syndrome; *NPV,* negative pressure ventilation.

Table II. Examples of diagnoses that lead to chronic respiratory failure and ventilatory dependence

Diagnostic group	Examples
Chronic pulmonary disease	Bronchopulmonary dysplasia
	Pulmonary hypoplasia
	Congenital heart disease
	Recurrent pneumonia/pneumonitis
Ventilatory muscle weakness	Duchenne muscular dystrophy
	Congenital myopathies
	Diaphragmatic dysfunction caused by phrenic nerve injury
	Thoracic dystrophies
Central hypoventilation syndromes	Congenital central hypoventilation syndrome
	Spinal muscle atrophy
	Cerebral/brainstem/spinal cord injury or malformation

Table V. Causes of death in children on chronic positive-pressure ventilation via tracheostomy reported in 13 previously published studies

Cause of death	Total n (%)
Progression of reason for CRF or other underlying condition	39 (34)
Unexplained/speculative	17 (14.5)
Infectious/sepsis/MODS	12 (10.5)
Acute respiratory failure	11 (9.5)
Congestive heart failure/cor pulmonale	11 (9.5)
Ventilator disconnection/airway accident	9 (8)
Seizures	3 (2.5)
Suicide	3 (2.5)
Arrhythmia	2 (2)
Hyperthermia	2 (2)
Post-operative cardiac complications	2 (2)
Head trauma/brain death	1 (1)
Post-operative hemorrhage	1 (1)
Ventilator failure	1 (1)
Total	114

Seven of these patients died after being weaned from chronic PPV, but the individual causes could not be extracted from the other reported deaths in the original paper.[3]

Table VI. Children's Hospital Los Angeles pre-discharge measures and supports for caregivers of children on home mechanical ventilation

Assessment/planning/services	•Discharge planning conferences with in-patient and out-patient pulmonology teams to address child's medical course and projected management •Assessment of the family's support system and psychosocial readiness for home care •Referral to community home care vendor to assess for home safety •Referrals for developmental, occupational, and/or physical therapy and schooling •All HMV patients qualify for 16 hours/day with licensed vocational nurse
Education	•At least two primary caregivers receive cardiopulmonary resuscitation and tracheostomy care training and must demonstrate independent competence before discharge •"Making Life Easier—Home Care Tips on Home Mechanical Ventilation," an educational DVD (available in both English and Spanish from Newport Medical Instruments at http://69.228.124.29/dvdvideo/hve.html) •"Home Care of Children on Ventilators: A Parent's Guide" (available online from the California Thoracic Society at www.thoracic.org/chapters/thoracic-society-chapters/ca/publications/resources/respiratory-disease-pediatric/Vent_Dependant_Children_booklet.pdf) •YouTube (LLC) informational video on managing children on HMV (found under "home mechanical ventilation") •Downloadable educational forms and lists are available
Equipment	•Pulse oximeter is provided for home as a back-up safety measure