Home mechanical ventilation: outcomes according to remoteness from health center and different family education levels

Sevgi Pekcan, Ayşe Tana Aslan, Nural Kiper, Mehmet Köse, Nazan Çobanoglu, Ebru Yalçın, Deniz Doğru, Uğur Özçelik

Pediatric Chest Diseases Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey

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Throughout the world, home mechanical ventilation (HMV) is being increasingly employed to treat patients suffering from chronic respiratory failure. This present study aimed to examine the characteristics and outcomes of 27 children seen in our department over a four-year period who were treated with HMV. The causes of chronic respiratory failure were as follows: 16 (59.3%) neuromuscular disease, 6 (22.2%) primary respiratory diseases, 3 (11.1%) congenital heart disease, and 2 (7.4%) storage disease. The mean age was 59.4 months (1 day-15 years); mean follow-up for invasive ventilation was 356 (0-1200) days and for non-invasive HMV was 517 (30-1440) days. With respect to maternal educational level, 13 had graduated from elementary school and 14 from high school or university. Nine of our patients resided in Ankara, while 18 lived in rural areas of Turkey. Eleven of the 27 patients died during the HMV period (1-36 months) at home. Five patients were weaned from HMV between 1-19 months.

Our experience showed that HMV can be applied successfully in chronic respiratory failure patients in Turkey. Length of the follow-up period and mortality rate were not affected by the patient's place of residence (city center or rural) or maternal level of education.

Key words: non-invasive home ventilation, invasive home ventilation, childhood, parental educational level, outcome.

The common etiologies of respiratory failure requiring ventilatory support in children include neuromuscular diseases, congenital central hypoventilation syndrome, spinal cord injury, craniofacial abnormalities, severe tracheobronchomalacia, chronic lung disease, and bronchopulmonary dysplasia[1,2]. The ventilation need of such children varies widely, from occasional use of continuous positive airway pressure (CPAP) by nasal mask to continuous positive pressure ventilation 24 hours a day through a tracheotomy tube. Therefore, carefully worked out care packages must be conceived on a case-by-case basis. Patient selection is the key to success for any home care ventilation program[3]. The goals of home mechanical ventilation (HMV) are: to enhance quality of life, to sustain and extend life without compromising quality, to improve or sustain physical and psychological function, to enhance growth and development, and to provide cost-effective care[4]. Since 1980, there has been an increase in the numbers of ventilator-dependent children and adolescents [5]. It is essential that a multi-professional, multi-disciplinary approach to their management be adopted from the outset[6]. While different patients will require different levels of care, it is hoped that each patient will progress to the point of maximum activity and take an active role in his or her own care. If this is accomplished, then the psychosocial well-being of the patient will also improve[7-10].

The aim of this study was to examine characteristics and outcomes of 27 pediatric patients seen in our institution over a four-year period who were treated with invasive or non-invasive HMV.

Material and Methods

Between January 2003 and January 2007, 27 children were diagnosed as pulmonary insufficiency in Hacettepe University Pediatric Chest Diseases Unit and were prescribed invasive (14 patients) or non-invasive (13 patients) HMV. All the patients with chronic respiratory failure who received HMV were included in the study.

Chronic respiratory failure implies that there is a chronic, perhaps irreversible, underlying respiratory disorder that causes respiratory insufficiency, resulting in inadequate ventilation or hypoxia. The primary indication for the use of HMV was chronic alveolar hypoventilation with associated respiratory failure indicated by hypoxemia and hypercapnia.

Patients were divided into two categories according to their residence in Ankara or outlying rural areas (minimum of 100 km from the city center). Fourteen patients were using pressure-controlled continuous ventilation, 12 continuous bi-level positive airway pressure (BiPAP) and 1 continuous CPAP.

The patients were thoroughly trained by doctors and respiratory therapists about using the home ventilatory equipment, aspiration, cardiopulmonary resuscitation, tracheostomy tube change, and postural drainage prior to the hospital discharge. A questionnaire evaluating the mother's age, educational level, socioeconomic status, and place of residence was given to all the parents.

We invited our patients for follow-up visits in a 2-3 month period. Those who were unable to attend were contacted by telephone to obtain the relevant information regarding the HMV treatment. Education on the use of equipment was reinforced during each visit. All patients and/or parents were queried regarding any problems with the set-up and control of the ventilator, their adaptation to the usage of the equipment, their preference for staying at home rather than at the hospital, and whether or not they accompanied their parents to places outside the home.

Statistical analysis was performed using SPSS, version 10.0. Mann–Whitney U test was used for statistical analysis. Values of $p<0.05$ were accepted as indicating significant differences.

Results

Twenty-seven patients participated in the study (19 male, 8 female; mean age: 59.4 months [1 day-15 years]). The mean follow-up period for patients with invasive mechanical ventilation was 356 (0-1200) days; for those with non-invasive mechanical ventilation the mean follow-up period was 517 (30-1440) days. Three patients had been hospitalized since birth. The other children were admitted after birth (38 days-19 years).

Six of the 14 patients who received invasive mechanical ventilation, the iVent 201 (Versamed) was used in 6, the LTV 900 (Viasys Healthcare) in 5 and the Carat II (Hoffrichter, Schwerin, Germany) in 3 patients. Eight of the 13 patients who received non-invasive mechanical ventilation used the BiPAP Harmony (Resipronic), 2 used the BiPAP Synchrony (Resipronic) and 3 used the Moritz ST (ResMed).

Eight children (61.5%) were on non-invasive mechanical ventilation via nasal mask while 5 (38.5%) used a face mask. Nineteen (70.3%) patients received ventilatory support for 24 hours and 8 (29.7%) were supported only during sleep. Twenty-five (92.6%) children received supplemental oxygen in addition to ventilatory support.

All patients' families were given training about HMV and rehabilitation care before

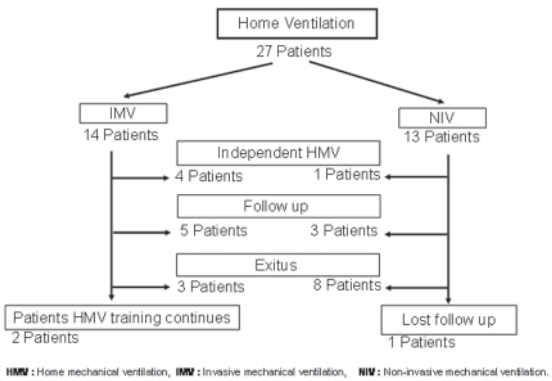

Fig. 1. Outcome of the patients.

Table I. Characteristics of Patients Administered Invasive Mechanical Ventilation

Gender (F/M) Residence (Ankara/ Rural)	Age at hospitalization /Age	Diagnosis	Duration of hospitalization	IMV application time	Duration of IMV application at home	Parents' education level Mother/Father	Outcome
F/ Rural	2 months/ 54 months	Operated CHD (VSD,PH,PS) MMR	407 days	407 days	510 days	Primary school/ High school	Independent of IMV
F /Rural	5 months/ 48 months	SMA	204 days	498 days	846 days	Primary school/ High school	Follow-up
F /Rural	108 months/ 114 months	CMD scoliosis	50 days	48 days	180 days	Primary school/ High school	Ex
F /Rural	45 days/ 30 months	SMA	23 days	13 days	660 days	Primary school Primary school	Follow-up
M/Ankara	6 months/ 55 months	SMA DE	269 days	259 days	1200 days	High school/ University	Follow-up
M /Rural	5 months/ 11 months	CHN	223 days	223 days	90 days	University/ University	Ex
M/Rural	4 months/ 10 months	HIE	115 days	115 days	90 days	University/ University	Ex
F/Ankara	1 day/ 18 months	CMD hydrocephalus	154 days	154 days	120 days	University/ University	Independent of IMV
M/Rural	2 days/ 22 months	Operated CHD (TOF, PDA, ASD, IPV)	144 days	144 days	600 days	University/ University	Independent of IMV
M/Rural	10 months/ 22 months	SMA	70 days	76 days	280 days	Primary school/ University	Follow-up
M/Ankara	24 months/ 32 months	MD	240 days	240 days	-	High school/ University	Training about HMV was continuing
M/Ankara	54 months/ 69 months	CMD	85 days		380 days	University/ University	Follow-up
M/Rural	2 days/ 16 months	Operated CHD (TAPVR) chylothorax	96 days	20 days	30 days	University/ University	Independent of IMV
F/Rural	38 days/ 5 months	MD	120 days	119 days	-	Primary school/ Primary school	Training about HMV was continuing

ASD: Atrial septal defect. **CHD:** Congenital heart disease. **CHN:** Congenital hypo-myelinization neuropathy. **CMD:** Congenital muscular dystrophy. **DE:** Diaphragm eventration. **Ex:** Exitus. **HIE:** Hypoxic-ischemic encephalopathy. **IMV:** Invasive mechanical ventilation. **IPV:** Imperforate pulmonary valve. **MD:** Mitochondrial disease. **MMR:** Mental motor retardation. **PDA:** Patent ductus arteriosus. **PH:** Pulmonary hypertension. **PS:** Pulmonary stenosis. **SMA:** Spinal muscular atrophy. **TAPVR:** Total anomalous pulmonary venous return. **TOF:** Tetralogy of Fallot. **VSD:** Ventricular septal defect. **M:** Male. **F:** Female.

discharge. The teams and departments involved in the follow-up of the patients included Pediatric Chest Diseases, Ear-Nose-Throat, Pediatric Cardiology, Physiotherapy, and the departments related with the primary illnesses. The causes of chronic respiratory failure were as follows: 16 (59.3%) neuromuscular disease, 6 (22.2%) primary respiratory diseases, 3 (11.1%) congenital heart disease, and 2 (7.4%) storage disease. Tables I and II present the diagnoses, age at hospital discharge, length of follow–up, outcome, family educational level, and place of residence. There were no life-threatening complications. Problems related to

Table II. Characteristics of Patients Administered Non-Invasive Mechanical Ventilation

Gender (F/M) Residence (Ankara/ Rural)	Diagnoses	NIV application age	Duration of NIV at home	Parents' education level Mother/ Father	Outcome
F/ Rural	Morquio S OSAS TI	144 months	12 months	University/ University	Exitus
M/ Rural	COPD	60 months	6 months	Primary school/ High school	Lost to follow-up
M Ankara	DMD VUR HT	85 months	32 months 3 hours/day	University/ High school	Follow-up
M/ Rural	OSAS	120 months before 72 days MV	48 months	University/ Primary school	Follow-up
M/ Rural	Hurler S PH MS	180 months	1 month	Primary school/ High school	Exitus
M/ Ankara	DMD scoliosis	144 months	9 months	University/ University	Exitus
F/ Rural	Encephalitis COPD scoliosis	141 months	36 months	Primary school/ High school	Exitus
M/ Rural	Cystic fibrosis Pectus carinatum	228 months	12 months	Primary school/ High school	Exitus
M/ Rural	ILD	162 month	9 months	Primary school/ High school	Exitus
F/ Ankara	SMA Scoliosis OSAS GERD	180 months	21 months	Primary school/ High school	Follow-up
M/ Ankara	Pycno-dysostosis Head injury	84 months	9 months	University/ High school	Exitus
M/ Ankara	Laryngeal dyskinesia Micrognathia dolicho-cephaly	3 months	16 months	University/ University	Independent of IMV
M/ Rural	SCID+BMT BO	57 months	13 months	Primary school/ Primary school	Exitus

BMT: Bone morrow transplantation. **BO:** Bronchiolitis obliterans. **COPD:** Chronic obstructive pulmonary disease. **DMD:** Duchenne muscular dystrophy. **GERD:** Gastroesophageal reflux disease. **HT:** Hypertension. **ILD:** Interstitial lung disease. **MS:** Mitral stenosis. **NIV:** Non-invasive mechanical ventilation. **OSAS:** Obstructive sleep apnea syndrome. **PH:** Pulmonary hypertension. **S:** Syndrome. **SCID:** Severe combined immune deficiency. **SMA:** Spinal muscular atrophy. **VUR:** Vesicoureteral reflux. **TI:** Tricuspid insufficiency. **F:** Female. **M:** Male.

ventilator equipment were seen rarely. Dermal laceration on the nasal bridge was seen in 2 patients. Oxygen concentrator failure was seen in 3 patients. Four of the 27 patients with tracheostomy reported accidental decannulation at home.

Eleven (40.7%) of the 27 patients died during the HMV period (1-36 months) at home. Figure 1 shows the patient outcomes. Death occurred within the first 7.5 months (1-36 months) following discharge. While 7 patients died at home, the other 4 patients died in the hospital. Diagnoses of the patients who died during the follow-up were neuromuscular disease (n: 6), pulmonary disease (n: 3) and storage disease (n: 2).

Table III. Comparison of the Follow-up and Exitus According to the Place of Residence of the Patients

	Ankara n: 9	Rural areas n: 18	
Follow-up period (days)	10-1200	10-1440	p>0.05
Exitus	2	9	p>0.05

At the end of the study period, 11 children remained ventilator-dependent. Five patients were ventilator-independent between 1-19 months, and their primary illnesses were congenital heart disease (n: 3), congenital muscular dystrophy (n: 1) and dyskinesia with micrognathia (n: 1).

With respect to maternal educational level, 13 (44.4%) mothers had graduated from elementary school, while 14 (55.5%) had graduated from high school or university. Nine (33%) patients resided in Ankara (location of hospital), while 18 (67%) resided in rural areas and/or suburban towns. There were no significant differences in the follow-up period or mortality rate according to maternal educational level or distance from our center (p>0.05) (Tables III, IV).

According to the results of the questionnaire, none of the mothers experienced any difficulties with the set-up or control of the ventilator equipment after receiving instruction and training in the hospital. All of the mothers reported their preference to remain at home rather than in the hospital. Nineteen families mentioned that the patients had accompanied them for all social activities outside the home, while eight families never took their patients outside the home due to the infection risks.

Discussion

Home mechanical ventilation for patients with chronic respiratory failure is an important, growing and successful technique for reducing morbidity and mortality, especially in those with chest wall and neuromuscular disease. HMV should be considered only when it is practical to provide the level of support required to meet the needs of the child and their family in the home[11]. The number of children dependent on long-term ventilatory assistance in Turkey, a developing country, is growing. Our experience has shown that in patients with chronic respiratory failure who come from different parts of the country and from families with varying education levels, both invasive and non-invasive mechanical ventilation can be applied successfully. The estimated prevalence of HMV was 6.6 per 100,000 people in the 16 European countries surveyed[3]. Home ventilator usage is very rare in childhood in our country, and there is currently very little experience in this regard. As the usage increases, more data and experience can be gained.

There are many benefits of home ventilation over continued ventilatory support in the hospital, including: reduction in the risk of nosocomial infection or in the development of antibiotic resistance; the opportunity to provide consistency in the caretakers, avoiding rotating teams or shifts of hospital personnel; the ability to match the level of ventilatory support with the usual activities of daily living; and the premise of a more familiar and relaxed environment with an improved quality of life [2,12,13].

Permanent location and residence of nine of our patients (one-third of the study group) was the city center (Ankara), while 18 of the patients resided in rural areas and/or suburban towns. Despite the disadvantages of rural habitation, HMV application was still successful. There were no significant differences in mortality rate or the follow-up period according to the

Table IV. Comparison of the Follow-up and Exitus According to the Mother's Education Level

	Mother's education Primary school n: 13	Mother's education High school n: 14	
Follow-up period (day)	10-1200	10-1440	p>0.05
Exitus	6	5	p>0.05

patient's place of residence.

We also surveyed the maternal educational level, since the mothers generally assume the real responsibility of the patient's care. The majority of the parents had completed primary school. Despite the low educational level of the mothers, they reported no difficulties with equipment applications or practice. There were no significant differences in mortality rate or the follow-up period according to the maternal educational level.

The most common underlying disease necessitating HMV in our cohort was neuromuscular disease (59.3%), and this is in accordance with previous literature[13-17]. The prognosis of long-term ventilation is probably related more to the underlying condition than to the mechanical ventilation[1]. Furthermore, mortality is also higher in patients with neurological diseases. In the present study, the mortality rate was 54%.

During the follow-up, five patients became completely ventilator-independent. The underlying disease in three of them was congenital cardiac disease. Need of ventilator was related to phrenic nerve injury during the surgical operation.

Discharge of the ventilator-dependent child requires a multidisciplinary approach and should be coordinated by one person responsible for liaising with all disciplines. Various teams and departments also contributed to and participated in our study. The departments involved in the patient follow-up included Pediatric Chest Disease, Ear-Nose-Throat, Cardiology, Physiotherapy, and any departments related with the primary disease[10].

Many other reports support the concept that home care is less expensive than hospital care[18]. Home care as an alternative to hospital care reduces costs by a reported 50-95%[19]. One home ventilator costs nearly 6,000 USD. Daily care in our hospital's intensive care unit is approximately 300 USD. The price of 20 days' care in our hospital approximately equals the cost of the home ventilator. The mean follow-up period was 163 days with HMV, which shows the benefit of HMV for our country's economy. In addition, the used device can be returned to the relevant social security system following death or when no longer needed.

Both invasive and non-invasive HMV enhanced the social life of the patients and facilitated their participation in outdoor life, benefitting both the patient and the family. Living at home rather than in the hospital improved the family's general level of happiness, psychology and well-being[13,20-23]. Quality of life is defined differently among patients and families. However, most families expressed satisfaction with an improved quality of life with home ventilation. A questionnaire evaluating life quality was not applied to the patient or the family, and this may be considered a limitation of the present study.

There were some social problems in the follow-up of these patients and their families. Currently, there are no professional home care nurses or respiratory therapists in Turkey. In the other countries like the United States and/or United Kingdom, nursing support is provided for the patients who use home ventilation, and special care facilities out of hospital are provided for the patients who can not be cared for at home for social reasons[24]. Oktem et al.[25], who studied 34 patients receiving ventilatory support at home, reported results similar to ours.

In conclusion, our experience has shown that HMV can be applied successfully in Turkey even when families reside in areas remote from the health care institution or when the educational level of the caregivers is low. We conclude that HMV in the pediatric age group plays an important role in developing countries and Turkey. Additionally, in view of its cost-effectiveness and easy application by parents with varying levels of education, it can be safely used in rural areas remote from the health center.

REFERENCES

1. Lloyd-Owen SJ, Donaldson GC, Ambrosino N, et al. Patterns of home mechanical ventilation use in Europe: results from the Eurovent survey. Eur Respir J 2005; 25: 1025-1031.

2. Srinivasan S, Doty SM, White TR, et al. Frequency, causes, and outcome of home ventilator failure. Chest 1998; 114: 363-367.

3. Tobin MJ. Advances in mechanical ventilation. N Engl J Med 2001; 344: 1986-1996.

4. Simonds AK. Home ventilation. Eur Respir J 2003; 47: 38s-46s.

5. Henderson J, Beckley S, England K, Booth W, Fleming P. Organizing ventilation at home. Curr Paediatr 2000; 10: 18-21.

6. Sritippayawan S, Kun SS, Keens TG, Davidson Ward SL. Initiation of home mechanical ventilation in children with neuromuscular diseases. J Pediatr 2003; 142: 481-485.

7. Fauroux B, Lofaso F. Non-invasive mechanical ventilation: when to start for what benefit? Thorax 2005; 60: 979-980.

8. Fauroux B, Boffa C, Desguerre I, Estournet B, Trang H. Long-term noninvasive mechanical ventilation for children at home: a national survey. Pediatr Pulmonol 2003; 35: 119-125.

9. Nava S. Non-invasive ventilation: a year in review. Breathe 2005; 1: 303-307.

10. Fiorenza D, Vitacca M, Clini E. Hospital monitoring, setting and training for home noninvasive ventilation. Monaldi Arch Chest Dis 2003; 59: 119-122.

11. Chu CM, Yu WC, Tam CM, Lam CW, Hui DS, Lai CK. Home mechanical ventilation in Hong Kong. Eur Respir J 2004; 23: 136-141.

12. Jardine E, Wallis C. Core guidelines for the discharge home of the child on long-term assisted ventilation in the United Kingdom. UK Working Party on Paediatric Long Term Ventilation. Thorax 1998; 53: 762-767.

13. DeWitt PK, Jansen MT, Ward SL, Keens TG. Obstacles to discharge of ventilator-assisted children from the hospital to home. Chest 1993; 103: 1560-1565.

14. Amin RS, Fitton CM. Tracheostomy and home ventilation in children. Semin Neonatol 2003; 8: 127-135.

15. Bertrand P, Fehlmann E, Lizama M, Holmgren N, Silva M, Sánchez I. [Home ventilatory assistance in Chilean children: 12 years' experience]. Arch Bronconeumol 2006; 42: 165-170.

16. Edwards EA, Hsiao K, Nixon GM. Paediatric home ventilatory support: the Auckland experience. J Paediatr Child Health 2005; 41: 652-658.

17. 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: 310-317.

18. Kurek CJ, Dewar D, Lambrinos J, Booth FV, Cohen IL. Clinical and economic outcome of mechanically ventilated patients in New York State during 1993: analysis of 10,473 cases under DRG 475. Chest 1998; 114: 214-222.

19. Frates RC Jr, Splaingard ML, Smith EO, Harrison GM. Outcome of home mechanical ventilation in children. J Pediatr 1985; 106: 850-856.

20. Goldstein RS, Psek JA, Gort EH. Home mechanical ventilation. Demographics and user perspectives. Chest 1995; 108: 1581-1586.

21. Adams AB, Whitman J, Marcy T. Surveys of long-term ventilatory support in Minnesota: 1986 and 1992. Chest 1993; 103: 1463-1469.

22. Carnevale FA, Alexander E, Davis M, Rennick J, Troini R. Daily living with distress and enrichment: the moral experience of families with ventilator-assisted children at home. Pediatrics 2006; 117: e48-60.

23. Jardine E, O'Toole M, Paton JY, Wallis C. Current status of long term ventilation of children in the United Kingdom: questionnaire survey. BMJ 1999; 318: 1699.

24. Hewitt-Taylor J. Children who require long-term ventilation: staff education and training. Intensive Crit Care Nurs 2004; 20: 93-102.

25. Oktem S, Ersu R, Uyan ZS, et al. Home ventilation for children with chronic respiratory failure in Istanbul. Respiration 2008; 76: 76-81.