

Long-Term Ventilation at Home and Pediatric Palliative Care: Patients' Characterization in an Italian Regional Survey

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Abstract

Background: The prevalence of children receiving long-term mechanical ventilation at home (LTMV-H) is rising in many countries as advances in technology and medicine extend the survival of such patients. In Italy, the prevalence is 4.2 per 100,000 pediatric patients. It becomes essential to thoroughly assess these patients' needs, and of the quality and adequacy of their home care. Pediatric palliative care (PPC) may be an appropriate strategy for the global care and quality of life of children needing LTMV-H and their families.

Objective: To characterize children on LTMV-H in the Veneto Region (north-east Italy) based on the experience of the Veneto Regional Center for PPC and Pain Control (VRCPPC).

Subjects: Children and adolescents (0-18 years of age) receiving invasive and noninvasive LTMV-H in the care of the VRCPPC from 01/09/2008 to 31/12/2013.

Methods: A retrospective cross-sectional analysis conducted by developing a regional database of children on LTMV-H.

Results: 56 children were on LTMV-H (mean age 4.5 years, 55% male), during the period considered. At the time of the survey, 38 of them were still in the care of the VRCPPC, 10 had died and 8 had been discharged. The children on LTMV-H accounted for 50% of all patients in the care of the VRCPPC (38/76 patients). They suffered mainly from neuromuscular diseases (NMDs) (17 patients), myopathy (11 pts), congenital central hypoventilation syndrome (CCHS) (7 pts), or nervous system disorders (7 pts). All patients had severe comorbidities. Their mean age when LTMV-H was started was 3.9 years. Ventilation was invasive (IMV) in 31 cases and noninvasive (NIV) in 25. Its initiation was mandatory in 39 cases (and in the intensive care setting for 36 of these children) and elective in 17 (and 15 of these children were in hospice care). Ventilation was provided for a mean 17.5 hours a days (14 for NIV; 18 for IMV). Median 9-year survival was 61%, but varied considerably by type of patient, being longer for NMDs, myopathy, CCHS, nervous system disorders (over 11 years in 78% of cases).

Conclusion: This population has complex life-long needs and numerous comorbidities that demand appropriate, continuous and qualified care. PPC could be an efficient strategy for meeting health goals and optimizing treatment planning.

Keywords: Long-term mechanical ventilation at home; Long-term home ventilation; Long-term ventilation, Home mechanical ventilation; Artificial ventilation; Chronic respiratory failure; Respiratory care at home; Pediatric palliative care; Life-limiting disease

Abbreviations:

HLTMV: Home Long-Term Mechanical Ventilation; PPC: Paediatric Palliative Care; VRCPPC: Veneto Regional Center for Paediatric Palliative Care and Pain Treatment; IMV: Invasive Mechanical Ventilation; NIV: Non-Invasive Ventilation; CCHS: Congenital Central Hypoventilation Syndrome; NMDs: Neuromuscular Diseases; OSAS: Obstruction Sleep Apnea Syndrome; ICU: Intensive Care Units; NICU: Neonatal Intensive Care Units

Introduction

In many countries the last two decades have seen a gradual increase in the prevalence of pediatric patients on long-term mechanical ventilation at home (LTMV-H) [1-12] as a result of advances in medicine and technology affording these children a longer survival [1-4,7-9,11,12]. This has given rise to a new pediatric population represented by young patients on LTMV-H, be it invasive (IMV) or non-invasive (NIV), that has been growing more rapidly in the last few years, especially the latter subgroup [4,13].

In Italy, the prevalence of pediatric patients on LTMV-H (both IMV and NIV) amounts to 4.2 per 100,000 pediatric patients. In 70% of cases, these patients are followed up by a team of pediatric specialists, pediatric intensive care operators, or pediatric pneumologists. The models of LTMV-H adopted around the country vary considerably,

and there is no national patient registry or shared protocol; this leads to important inequalities in the distribution and quality of this treatment on a national scale [5,6].

Despite the limited numbers involved, this population engenders high costs for the health services [1,2,7,14], partly as a consequence of the severe comorbidities usually involved [1,3] that make it necessary to involve multiple specialists, services and institutions to provide adequate social support and health care.

Like all children with incurable chronic diseases, the ideal place to care for these patients is in their own homes, as unanimously confirmed by the literature [15-23]. In recent years, technological advances have enabled the use of mechanical ventilation at home, but home care for a child needing mechanical ventilatory support demands the adoption of a highly complex patient care plan [7-14]. This includes drawing up a detailed program for providing medical and nursing care, and organizational support [7,14,22-24], as well as planning adequate training for family caregivers and territorial healthcare operators [25], a patient monitoring schedule, procedures for managing emergencies and, in due course, appropriate arrangements for handing the patient over to the adult services [26-28].

In December 2003, the Veneto Regional Authority was the first in Italy to promote and organize the setup of a regional center for pediatric palliative care and pain control (the VRCPPC), which serves as a clinical, organizational and scientific reference for the whole region inasmuch as concerns the management of pediatric patients with pain and/or incurable disease.

Patients on LTMV-H account for a significant proportion of the population offered pediatric palliative care (PPC), that is estimated in the literature to be in the range of 50-90% [1-3,29,30].

Objective

The aim of this study was to identify the demographic and clinical features of the pediatric patients on LTMV-H in our region [1-8,10-13,31-35] with a view to making the best possible arrangements for their care.

Materials and Methods

This study was a retrospective cross-sectional analysis of all pediatric patients on LTMV-H taken into care by the VRCPPC between 1 September 2008 and 31 December 2013. These patients on LTMV-H were defined as clinically stable cases needing daily invasive (IMV) or noninvasive (NIV) ventilatory support for at least three months who were managed at home and/or in a hospice setting [2-4,7]. This included all patients on LTMV-H taken into care by the VRCPPC at some point during the period considered, both those still in the center's care at the time of the study and those who had died or

been discharged. Patients who were tracheostomized, and who had therefore been ventilated previously, but were not on LTMV-H when they were in the care of the VRCPPC, were excluded from the study sample.

Our analysis focused on the demographic and clinical characteristics of the study sample, including: age and gender, principal diagnosis, comorbidities, the physician referring the patient to the VRCPPC, the amount of time spent in the center's care, the place where ventilation was initiated, the reason and whether the treatment was mandatory or elective, the type of ventilation (IMV vs. NIV), the patient's age when ventilation was started, the reason for any tracheotomy and the patient's age at the time of this procedure, the duration of ventilatory support (hours per day), the patient's age when ventilation was suspended and a tracheostomy was closed. We developed a regional database in which we recorded all the variables considered. The sources of information considered were patients' clinical records (on printed matter and electronic media), and the computer-based home care records collected by the VRCPPC for all patients in the center's care. The descriptive analysis included absolute and relative frequencies of the quantitative variables, means \pm standard deviations for normally distributed quantitative variables, and the main quartiles (medians, first and third quartiles) in the case of quantitative variables that violated the assumption of a normal distribution (Shapiro-Wilk test). For the between-group comparisons, the inferential tests used were the t-test for quantitative variables satisfying the assumption of a normal distribution, the Wilcoxon-Mann-Whitney test for quantitative variables with a non-normal distribution, the log rank test for the survival analysis, and the chi-square test for assessing the independent quantitative variable hypothesis. In all the tests significance was assumed for probability values below 0.05. The statistical analyses were conducted with the R software rel. 3.0.1 and Microsoft Excel 2007.

The study has obtained the approval by the Ethics Committee and patients and their families have given their informed consensus.

Results

There were 56 pediatric patients on LTMV-H in the care of the VRCPPC during the period considered; 55% of them (31/56 patients) were male, and the mean age of the sample was 4.5 years (SD 4.9). The principal diagnosis is showed in Table 1.

All patients had severe comorbidities, mainly of neuromotor (96%) and osteoarticular (71%) type, but also cardiological (36%), nephrological (20%) and neurocognitive (32%). There were also sensory (visual and/or hearing) impairments in 23% of cases.

The place and the mean age of starting ventilation in relation with the main diagnosis are showed in table, together with the modality of starting ventilation (NIV or IMV) and the duration of ventilatory support (hour/day) (Table 1).

Principal diagnosis (number of patients)	Place of starting ventilation	Age of starting ventilation (yrs)	Modality of ventilation	Mean duration of ventilation (hour/day)
Heart disease (4 patients)	ICU/NICU 4 pts	0.4 (\pm 1.8)	IMV 4 pts	19.5 h
Central hypoventilation (7 pts)	ICU/NICU 7 pts	1 (\pm 1.8)	NIV 1 pts IMV 6 pts	12 h if IMV 8 h if NIV

Metabolic disorders (3 pts)	ICU 3 pts	1.4 (±0.8)	IMV 3 pts	24 h
Other disorders (5 pts)	ICU/NICU 5 pts	2.3 (±4.2)	IMV 5 pts	18 h
Neuromuscular disorders (17 pts)	ICU 8 pts Hospice 9 pts	4.1 (±4)	NIV 15 pts IMV 2 pts	19.5 h if IMV 12 h (±5) if NIV
Myopathy (11 pts)	ICU/NICU 7 pts Hospice/ward 4 pts	5.75 (±6)	NIV 6 pts IMV 5 pts	18 h if IMV 12 h (±5) if NIV
Nervous system disorders (7 pts)	ICU 4 pts Hospice 3 pts	6.3 (±6.8)	NIV 3 pts IMV 4 pts	20 h if IMV 7 h (±5) if NIV
Oncohematological diseases (2 pts)	ICU 2 pts	8.5 (±10.5)	IMV 2 pts	14.5 h if IMV
56 pts	ICU/NICU 40 pts Hospice 16 pts	3.9 (±5)	NIV 25 pts IMV 31 pts	18 h (±6) if IMV 14 h (±7) if NIV 17.5 h (±5)

Table 1: Main diagnosis relating with place and mean age of starting ventilation, modality and duration of ventilation.

The provision of ventilatory support in acute conditions was followed by a tracheotomy in 72% of cases and this procedure was implemented in 75% of the children started on ventilation in an intensive care setting. All the tracheostomies were surgically performed. The most common reason for tracheotomy was prolonged intubation in patients impossible to extubate (80% of cases); other causes were frequent intubations (7%), elective indications in relation to the principal diagnosis (7%), bedsores and episodes of severe apnea. The main patient's age at the time of the procedure, relating with the main diagnosis, is showed in Table 2.

Principal diagnosis (number of patients)	Age of tracheotomy (yrs)
Heart disease (4 patients)	0.4 (+0.4)
Central hypoventilation (6 pts)	0.6 (+1)
Metabolic disorders (3 pts)	1.9 (+0.9)
Other disorders (5 pts)	2.4 (+4.2)
Neuromuscular disorders (2 pts)	0.7 (+0)
Myopathy (5 pts)	2.2 (+4)
Nervous system disorders (4 pts)	3 (+3.9)
Oncohematological diseases (2 pts)	4.25 (+4.5)
31 pts	1.8 (+2.9)

Table 2: Mean age of tracheotomy in relation with the main diagnosis.

Monitoring infectious diseases was performed during each valuation of FU (about every 4-6 months in relation to the clinical situation) and whenever the child showed symptoms and/or signs of respiratory infection. None of the patients in NIV was colonized. The % of patients tracheostomized colonized is high: 70%. The pathogen most frequent involved is the *Pseudomonas aeruginosa* (others are *P. mirabilis* and *A. baumannii*).

Ventilation was suspended in 4 patients on LTMV-H (one with heart disease, one with a nervous system disorder, and two with other conditions). The mean age of these patients whose ventilation was suspended was 1.2 years (SD 0.7).

The tracheotomy was closed in 3 patients and the patient's age at the time was 16 years in one case, and within the first two years of life in the other two (at 2.2 and 1.6 years old).

Considering the sample as a whole, 38/56 patients were still in the care of the VRCPPC at the time of our study, while 10/56 had died and 8/56 had been discharged. These 38 patients on LTMV-H accounted for 50% of all the patients in the care of the VRCPPC as at 31 December 2013 (38/76 patients), for 10.3% of the patients taken into care by the center who died (10/97 patients), and for 6.6% of those who were discharged (8/121 patients). The number of ventilated patients in the care of the VRCPPC increased annually by approximately 10%.

The 9-year survival rate of patients on LTMV-H was 61%, with a marked variability depending on their diagnosis. Among the patients with NMDs and myopathy, CCHS and nervous system disorders, 78% were still alive at 11 years, while the patients with heart disease, metabolic disorders, oncohematological and other diseases did not survive beyond 9 years (p 0.041).

Discussion

Patients on LTMV-H accounted for 19% of all patients taken into care by the VRCPPC during the period considered, consistently with other reports in the literature [29,30]. When only the patients still in the care of the center at the time of our study were considered, those on LTMV-H represented 50% of the sample (given the open nature of the cohort that gives rise to a cumulative prevalence).

The mean age of the patients on LTMV-H in the care of the VRCPPC was 4.5 years (range 0-18 years). Unlike the series described in other studies [29,30], there were no older cases because patients are transferred to the adult services when they reach the age of 18, in accordance with current Italian legislation. The mean age of our

sample was much lower than in other studies concerning pediatric populations on LTMV-H [1-8,10-12,31-35]: the Canadian studies [3] describe a sample with a mean age of 9.5 years, and the other Italian reports concern samples with a mean age of 8 years [6]. As for gender, the distribution in our cohort revealed no major differences among patients requiring LTMV-H, with only a slight prevalence of male gender (55%), as reported elsewhere [13].

The percentage distribution of the various diseases and disorders in our sample was only partially consistent with other reports [1-8,10-13,31-36]: while the percentages relating to NMDs and CCHS were similar, there was a much lower rate of lung and airway diseases (bronchial dysplasia, malaria, ciliary dyskinesia, OSAS, and other respiratory diseases with chronic hypoventilation) in our sample, with 3.6% as opposed to 25-35% in the literature [1-6,8,9,11-13,31-36]. This may relate to patient care and clinical or organizational factors particular to our setting: patients are only referred to the regional reference center if they are under 18 years old and have complex, life-threatening diseases posing very complicated care issues. As reported elsewhere in the literature [1-3,13,37], our patients all had severe comorbidities, whereas their frequency was lower in the samples described in British studies (67%) [2,37].

The age at the time of starting ventilator support was a mean 3.9 years; it was lower in children whose ventilation was initiated in acute conditions (2.6 years), and higher for those whose ventilation was started electively (7 years). By comparison with other reports in the literature, our patients were very young when they started on ventilatory support (the Canadian studies reported a mean age of onset of 9.6 years), though the distribution by type of ventilation adopted was consistent with the literature [3,6,32], i.e. at 10.6 years old in the case of NIV, and 1.1 years of age in the case of IMV. There are currently no data in the literature with which to compare the age at the time of starting ventilation in relation to main diagnosis.

When ventilatory support was started, it was invasive (IMV) in 55% of cases, involving the need for a tracheotomy. For 45% of the patients, ventilatory support was initially noninvasive type (NIV). There are currently no data available in the literature with which to compare the type of ventilation (NIV vs. IMV) in relation to the patients' principal diagnosis.

Tracheotomies were performed mainly in acute conditions (72%) and in the majority of children placed on ventilatory support in an intensive care department (75%). In 80% of cases the procedure became necessary after prolonged intubation in patients impossible to extubate [37], and generally at an early age (mean 1.8 years). These findings are consistent with the content of the national [5,6] and international [3,13,36] literature.

A duration of ventilation averaging 17.5 hours a day is also consistent with other reports [5]. Consistently with the international literature [3], ventilatory support was provided for shorter mean periods in patients on NIV (mean 14 hours, minimum 7 hours), which generally did not exceed the 16-hour cut-off mentioned in the literature as an indication for tracheostomy [36].

Ventilatory support was suspended in only 7% of our patients on LTMV-H, at a mean age of 2.4 years. The percentage of our patients on LTMV-H who could be weaned from the ventilator is consistent with some reports in the literature (6.6%) [1], but lower than in others (19-21%) [4,13]. These differences may be due to our sample including fewer patients on LTMV-H for respiratory diseases, whose condition generally improves as the lung develops during the child's physiological

growth, often enabling such patients to be completely weaned from the ventilator. This is confirmed by the fact that tracheostomies were closed in 10% of the patients who underwent this procedure, and mainly in patients whose hypoventilation was due to the effects of extreme prematurity.

Among the patients forming the object of this study, 64% were still in the care of our center. This figure indicates the cumulative prevalence due to the open nature of the sample, and reflects an increase in the incidence and prevalence of patients on LTMV-H in the pediatric population, as already reported elsewhere [1-4,7-9,11,12]. On the other hand, 18% of our sample concerned patients who died, a higher percentage than in some other reports, e.g. 10% in the British report [1] and 15% in the Canadian ones [3]. These differences may relate to the monocentric nature of our study and consequently small size of our sample by comparison with the multicentric British and Canadian reports. On the other hand, the mortality rate was far lower in our sample than in reports on pediatric patients receiving PPC (30% at 1 year) [30]; the difference probably relates to the oncohematological patients, who were scarcely represented in our sample. Consistently with other reports on patients receiving PPC [29-30], 14% of the patients in our sample were discharged. Clearly, given the gradual improvement in the survival rates for this population [1-4,13,30], and the consequent increase in the number of patients reaching adulthood (26-28% in the Canadian studies) [3,4], we need to develop dedicated pathways for the patients' transition from PPC to the adult services [26-28]. The approximately 10% annual increase in the number of patients on LTMV-H in the care of the VRCPPC during the period considered confirms the constant increase in the prevalence of this population already reported in the literature [1-4,7-9,11,12,29,30].

The overall 9-year survival rate in our sample was 61%, lower than in another study reporting 5- and 10-year survival rates of 94% and 91%, respectively [4]. If we consider only the patients with NMDs, myopathy and CCHS, the survival rate is more similar to the international datas.

Conclusions

The population considered in this study was consistent with the literature on pediatric populations on LTMV-H. Our findings only concern the situation in the Veneto region and need to be extended to the whole of Italy in the near future. Creating a database that pools information from all the other regions in the country too and establishing a national register of pediatric patients on LTMV-H would generate an indispensable knowledge source on which to base adequate, competent and equitable organizational solutions that meet the real needs of this particular population.

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