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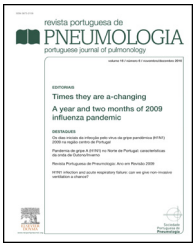

ORIGINAL ARTICLE

Long-term ventilation in children: Ten years later

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Abstract
Introduction: Home mechanical ventilation (HMV) represents a treatment option for patients with chronic respiratory failure and has changed prognosis and survival of many disorders in children. The aim of this study was to characterize a group of children on long-term mechanical ventilation (LTMV) for a period longer than 10 years.
Methods: A retrospective analysis was carried out including patients on LTMV for more than 10 years (LTMV-10) in a tertiary pediatric hospital. Statistical analysis: PASW Statistics 18®.
Results: Thirty-one children (61% female) belong to the LTMV-10 group. Median age at the beginning of ventilatory support was 3 years (birth to 13 years). Main indications for assisted ventilation were neuromuscular disease ($n = 12$, 39%), metabolic disease ($n = 7$, 23%) and central hypoventilation ($n = 6$, 19%). Volume ventilation was used in 2 children, and positive pressure ventilation in the others, mainly bilevel positive airway pressure ($n = 25$, 81%). Invasive ventilation via tracheostomy was used since the beginning in four cases, and subsequently in two other children. The mean time of ventilatory support was 146 months and the maximum was 219 months. Respiratory morbidity was the most frequent cause of hospitalization and the annual rate of such episodes was 0.17 per child. Global mortality rate was 19%.
Conclusions: HMV programs provide necessary and safe assistance for children with severe chronic respiratory failure. As shown in our series, it is possible to be kept on this respiratory support modality for long periods with good compliance and a small number of hospitalizations.

Introduction

In the last two decades there has been an increase of children requiring long-term mechanical ventilation (LTMV) and, in many countries, programs of home mechanical ventilation (HMV) have been developed.[1–6] Several factors have contributed to this: advances in neonatal and

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pediatric care with improved survival of children with acute respiratory insufficiency, technological advances and development of suitable home equipment, and increased use of polysomnography (PSG) to diagnose sleep disordered breathing.[5,7] Furthermore, pressure to reduce hospital stay duration and awareness that this environment is inappropriate for children, has resulted in increased number of patients being discharged on ventilatory support.[8]

In this context, HMV, mainly non-invasive ventilation (NIV), provides domiciliary health care for patients with chronic respiratory failure. HMV combines significant change in family life, increased survival rate, quality of life improvement and decrease in hospital costs.[3,7,9–11]

In 1993, a HMV program for patients with chronic respiratory failure began in our hospital, with a multidisciplinary team formed by pediatric pulmonology physicians and nurses, psychologists and social workers. In the last 20 years, more than 300 children have been supported with HMV.

The aim of this study was to characterize the group of children and adolescents on HMV for a period longer than 10 years followed in our hospital and analyze compliance and impact of HMV. Patients on LTMV transferred to other institutions before a 10 years of follow-up, were excluded, although some may have kept ventilatory support.

Materials and methods

An observational study was performed through data collection and retrospective review of medical records of pediatric patients on HMV for longer than 10 years (LTMV-10) followed in our hospital. Starting point for follow-up was day one of ventilatory support and end point was the moment of data collection conclusion (31st August 2013), patient's transferal or death.

Data collected included patient's age, gender, diagnosis, age at beginning of ventilatory support, indications for its use, ventilatory technique and mode of ventilation provided, previous overnight oximetry or PSG, length of time on HMV, patient's compliance and outcome.

Data were collected and analyzed through PASW Statistics 18®.

Results

Since 1993, 31 children were under HMV for a period longer than 10 years and were followed in our hospital. Nine patients (29%) started HMV between 1993 and 1998 and 22 (71%) between 1999 and 2003 (Fig. 1). The patients were categorized according to diagnosis: neuromuscular disorders (NMD) – 12 (39%), metabolic diseases – 7 (23%), central hypoventilation – 6 (19%), genetic syndromes – 2 (6%) and other neurological disorders – 4 (13%) (Table 1). Nineteen patients (61%) were female. Gender was equally distributed among all diagnostic groups.

The average age of beginning ventilatory support was three years, ranging from birth to 13 years depending on the diagnosis (Fig. 2). Congenital central hypoventilation syndrome (CCHS) children were subsequently on NIV, starting between 11 days and 5 months of age, with the exception of one case that stayed on invasive ventilation (tracheostomy) since birth.

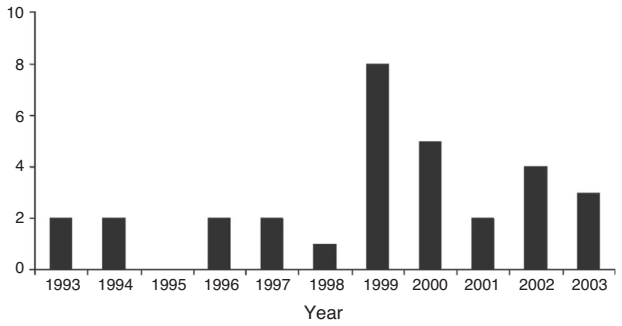

Figure 1 Number of LTMV-10 children initiated on respiratory support per year.

Ventilatory support was started electively in 17 (55%) children (Table 2). In 12 patients, this decision was supported by a full PSG or nocturnal oximetry: PSG showed hypoventilation in 3 congenital muscular dystrophy (CMD), 1 congenital myopathy and 1 mucopolysaccharidosis (MPS) and obstructive sleep apnea (OSA) in 2 MPS, 1 Duchenne muscular dystrophy (DMD) and 1 corpus callosum agenesis; recurrent desaturations on oximetry in 1 CMD, 1 Prader-Willi syndrome (PWS) and 1 glycogen storage disease (GSD) type II. In three children with spinal muscular atrophy (SMA) type II, ventilatory support was started in the first two years of life on a prophylactic basis. Two other patients (CMD and SMA type II) started ventilatory support due to clinical symptoms of hypoventilation.

Fourteen patients (45%) had ventilatory support initiated in emergency situations. Six started in the first days of life (Table 1) and could not be weaned from ventilation and four (2 MPS, 1 NMD and 1 GSD type II) began during an acute respiratory illness. The remaining encloses one PWS with hypercapnic coma, one neurodegenerative disease requiring tracheostomy and two previously healthy children (sequelae of pneumococcal meningitis and cervical spinal cord trauma).

LTMV was started mainly by NIV in most children ($n = 27$, 87%). Invasive ventilation through tracheostomy was initiated in four children (Fig. 3). During follow-up, two more patients required tracheostomy: one MPS with irreversible airway obstruction and one CMD before surgery for scoliosis.

Seven children (five on NIV and the two with tracheostomy) needed to increase the daily time of ventilation to 16–20 h or full day.

Volume ventilation was used in two of the first children included and positive pressure ventilation in all the other cases, mainly bilevel positive airway pressure (BPAP) – 81% (Table 2). Continuous positive airway pressure (CPAP) was used in the four children with OSA. Three patients (1 DMD and 2 MPS) needed a subsequent change of mode of ventilation to BPAP due to association of alveolar hypoventilation.

Other support equipment was provided according to patient' needs: oxygen saturation monitor, humidification system, suction device, mechanical in-exsufflator and self-inflating resuscitation bag.

During the last five years of follow-up, 8 (26%) patients had 5–10 respiratory infections, 12 (39%) had less than 5 and