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► **To cite this version:**

Fabrizio Racca, Giacomo Berta, Marco Sequi, Elisabetta Bignamini, Elena Capello, et al.. Long-term Home Ventilation of Children in Italy: a National Survey. *Pediatric Pulmonology*, 2010, 46 (6), pp.566. 10.1002/ppul.21401 . hal-00613787

**HAL Id: hal-00613787**

**<https://hal.science/hal-00613787>**

Submitted on 6 Aug 2011

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**Long-term Home Ventilation of Children in Italy: a National Survey**



Journal:	<i>Pediatric Pulmonology</i>
Manuscript ID:	PPUL-10-0162.R2
Wiley - Manuscript type:	Original Article
Date Submitted by the Author:	10-Sep-2010
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Keywords:	Chronic respiratory failure, home respiratory care, epidemiology, disability, long term ventilation, home mechanical ventilation

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 Manuscripts

## Long-term Home Ventilation of Children in Italy: a National Survey

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**Funding:** none

**Short title:** Home Mechanical Ventilation for Italian Children

1  
2  
3 **Keywords:** Chronic respiratory failure, home respiratory care, long term ventilation, home  
4 mechanical ventilation, epidemiology, artificial ventilation, disability.  
5

6  
7 **Word count:** 3019

**Number of tables:** 1

8  
9 **Number of figures:** 0

**Conflicts of interest:** none.  
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59  
60**ABSTRACT**

**Background:** Improved technology, as well as professional and parental awareness, enable many ventilator-dependent children to live at home. However, the profile of this growing population, the quality and adequacy of home care, and patients' needs still require thorough assessment.

**Objectives:** To define the characteristics of Italian children receiving long-term home mechanical ventilation (HMV) in Italy.

**Methods:** A detailed questionnaire was sent to 302 National Health Service hospitals potentially involved in the care of HVM in children (aged < 17 yrs). Information was collected on patient characteristics, type of ventilation, and home respiratory care.

**Results:** A total of 362 HMV children was identified. The prevalence was 4.2 per 100,000 (95% CI: 3.8-4.6), median age was 8 years (interquartile range 4-14), median age at starting mechanical ventilation was 4 years (1-11), and 56% were male. The most frequent diagnostic categories were neuromuscular disorders (49%), lung and upper respiratory tract diseases (18%), hypoxic (ischemic) encephalopathy (13%), and abnormal ventilation control (12%). Medical professionals with nurses (for 62 % of children) and physiotherapists (20 %) participated in the patients' discharge from hospital, though parents were the primary care giver and in 47% of cases, the sole care giver. Invasive ventilation was used in 41% and was significantly related to young age, southern regional residence, longer time spent under mechanical ventilation, neuromuscular disorders, or hypoxic (ischemic) encephalopathy.

**Conclusions:** Care and technical assistance of long-term HMV children need assessment, planning, and resources. A wide variability in pattern of HMV was found throughout Italy. An Italian national ventilation program, as well as a national registry, could be useful in improving the care of these often critically ill children.

## INTRODUCTION

Continuous advances in neonatal and pediatric intensive care are likely to result in more infants, children, and adolescents with critical, chronic conditions, who survive and who are discharged home, but require long-term technological support.<sup>1-3</sup> A growing population of children exists who has chronic respiratory failure due to conditions such as neuromuscular diseases, obstructive sleep apnea, craniofacial abnormalities, or spinal cord injuries, and who require either tracheostomy or mask ventilation.

Pressure to reduce the hospital stay of these children and awareness of the effects of long term hospitalization on a child's development have, during the last decade - especially, in a few countries such as UK, France, and US, - increased the numbers of ventilated children discharged home. However, the availability and practical details of complex respiratory support at home involve a lot of variables such as appropriate home environment, healthcare support, and funding, that cannot always be guaranteed. Findings of the Eurovent study, a wide cross sectional survey including adults as well as children, highlighted how the collection of information about home mechanical ventilation (HMV) could facilitate national planning for HMV. In particular, it could aid the development of HMV services in order to ensure equality of provision and access.<sup>3</sup>

Surveys in Canada,<sup>4,5</sup> Chile,<sup>6</sup> Germany,<sup>7</sup> France,<sup>8,9</sup> Italy,<sup>10,11</sup> Japan,<sup>12</sup> New Zealand,<sup>13</sup> Switzerland,<sup>14</sup> The Netherlands,<sup>15</sup> Turkey,<sup>16</sup> UK,<sup>17-19</sup> and USA<sup>20-22</sup> have defined the size, main features, and needs of the pediatric population requiring home mechanical ventilation. However there are many differences between these observational studies: the setting (single referring center, state, or country), the time period (one year or trend over time), the data collected, and how it was collected. Thus, estimates of the national prevalence of home ventilator-dependent children range from 0.7/100,000 children in 1993 in Japan<sup>12</sup> to 3.4/100,000 in 2000 in Switzerland.<sup>14</sup>

Because information on long-term home ventilation for children in Italy is scant and erratic (no regional or national population-based data exist), this survey was conducted to describe the size and

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2  
3 characteristics of the pediatric Italian population receiving chronic mechanical ventilation at home  
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5 and the profile of home respiratory care.  
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## 10 **METHODS**

### 11 **Study Population**

12  
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14 A questionnaire was sent to all 27 Italian pediatric intensive care and pulmonary units to get data on  
15  
16 children up to 17 years old receiving HMV in January 2007. To identify as many children as  
17  
18 possible the questionnaire was sent also to another 275 hospital-based units (eg. pulmonary  
19  
20 services, and critical care units) of the National Health Service (NHS) that are potentially involved  
21  
22 in the care of the target population. Information referred to children discharged home from hospital  
23  
24 where required tracheostomy or mask ventilation.  
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### 29 **Home respiratory care**

30  
31 HMV was defined as invasive mechanical ventilation (IMV) or non invasive mechanical  
32  
33 ventilation (NIV), for a period of at least 3 months on a daily basis, carried out mostly in the user's  
34  
35 home or other long-term care facility (not a hospital).<sup>3</sup> Basic demographic data (age, sex, place of  
36  
37 residence), causes of respiratory failure, hospitalization history, duration of ventilatory support, type  
38  
39 of ventilation and management-related description and home care provider information were  
40  
41 collected.  
42  
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### 46 **Data analysis**

47  
48 Data were entered into a SAS/STAT database (Statistical Analysis Software, Cary, NC) for  
49  
50 recording and analysis. The prevalence with 95% confidence intervals (CI) of Italian children  
51  
52 ventilated at home on the long term was calculated using the Istat 2007 annual report (Italian  
53  
54 National Institute of Statistics) for children up to 17 years old, resident in the provinces in which  
55  
56 patients were identified as the denominator. Age was calculated on the basis of the date of birth and  
57  
58 the date of onset of ventilation was recorded.  
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3 Descriptive statistics were computed for the entire study population and subgroups. Student's *t* test  
4 was used to compare the type of ventilation and patient's age (continuous variable).  $\chi^2$  tests were  
5 used to compare the type of ventilation and categorical variables, such as sex and home health care.  
6  
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9  
10 The Mann-Whitney U test was used to compare the type of ventilation in each class of time/day  
11 spent under mechanical ventilation. ANOVA was used to check the association between the type of  
12 ventilation and class of diagnosis. Comparisons of the type of ventilation in each region were tested  
13  
14  
15 by  $\chi^2_{\text{trend}}$  analysis. Multivariate logistic regression was used to look at the association between the  
16  
17  
18 type of ventilation and all monitored factors, simultaneously.  
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21  
22 Results are presented as frequencies, or median and interquartile range;  $P < 0.05$  was considered  
23  
24 significant.  
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26  
27 All data collected on centres and patients were kept strictly confidential. Data on patients were  
28  
29 anonymous. The San Giovanni Battista-Molinette Hospital, University of Turin, Committee on  
30  
31 Clinical Investigation reviewed and approved the study protocol. Because this was an observational  
32  
33 study without an intervention and without the presentation of any identifying data, the committee  
34  
35 ruled that informed consent was not necessary.  
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## 41 RESULTS

42  
43 A total of 362 children were identified by 30 centers (67% in the North of Italy) as receiving home  
44  
45 mechanical ventilation in January 2007, with an estimated prevalence of 4.2 per 100,000  $\leq 17$  year  
46  
47 old children (95% CI 3.8-4.6). Patients were resident in 75 of the 107 Italian provinces (70%); 200  
48  
49 (55%) were resident in the North, 55 (15%) in the Center, and 107 (30%) in the South, in good  
50  
51 agreement with the national distribution of children up to 17 years old. No difference was observed  
52  
53 between the type of ventilation and the area of residence (Table 1). The majority of patients (59%)  
54  
55 were discharged from 12 of 27 NHS pediatric intensive care units or pediatric pulmonary units that  
56  
57  
58 were following mechanically ventilated children at home, and the others came from 18 other  
59  
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3 hospital units and wards (i.e., adult intensive care units, adult pulmonary units, pediatric neurology  
4  
5 units, pediatric palliative care units). The median number of children followed by each center was  
6  
7 seven (interquartile range 3-19).

8  
9  
10 The median age was 8 years (interquartile range 4-14) and was significantly different for children  
11  
12 receiving invasive or non-invasive ventilation ( $P \ll 0.001$ ), as well as for the median age at the  
13  
14 start of mechanical ventilation ( $P \ll 0.001$ ) (Table 1). All children received positive pressure  
15  
16 ventilation. The majority of children receives NIV (Table 1), which is delivered mostly by nasal  
17  
18 mask (85% of patients receiving NIV). Non-invasive ventilator support was the most used and  
19  
20 skewed toward late infancy and adolescence.. Overall, 41% of the survey population had ventilation  
21  
22 via tracheotomy with the highest percentage of children affected by neurological disorders. The  
23  
24 median duration of home mechanical ventilation was one year (Table 1).  
25  
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28  
29 The most frequent diagnostic category was neuromuscular disorders (49%), followed by lung and  
30  
31 upper respiratory diseases (18%), hypoxic (ischemic) encephalopathy (13%), and abnormal  
32  
33 ventilation control (12%) (Table 1). The most represented neuromuscular disorders were spinal  
34  
35 muscular atrophy (SMA) (78 patients), Duchenne muscular dystrophy (31 patients) and Congenital  
36  
37 muscular dystrophy (29 patients). In particular, 25 children were affected by SMA type 1 (16  
38  
39 receiving NIV, 64%) and 53 children by SMA type 2 (30 receiving NIV, 57%).  
40  
41  
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43 There were wide difference between the type of ventilation and diagnoses (  $F = 5.94, P < 0.016$ ).

44  
45 Time per day spent under mechanical ventilation was longer for children requiring invasive  
46  
47 ventilation ( $\chi^2 = 148; P \ll 0.001$ ) (Table 1), particularly for those with neuromuscular disorders ( $\chi^2$   
48  
49  $= 73; P \ll 0.001$ ), hypoxic (ischemic) encephalopathy ( $\chi^2 = 37; P \ll 0.001$ ), or chronic lung and  
50  
51 upper respiratory diseases ( $\chi^2 = 28; P \ll 0.001$ ) .  
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54  
55 Nurses (for 62 % of children), physiotherapists (20 %), social workers (7 %), psychologists (7 %),  
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57 pediatricians (5%), and other professionals (4%) participated in the home discharge process with  
58  
59 hospital physicians. All children were involved in a patient ventilator training program (94% during  
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3 inpatient visits; 6% during outpatient visits) and in a regular monitoring program. At home visits  
4  
5 were planned in 60% of patients (36% of patients only during planned hospitalization, 30% only  
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7 during outpatient visits, 29% with both modalities, 5 and % only at home). The mean number of  
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9 hospital visits was 2.8/year, while the mean number of at home visits was 27/year. The at home visit  
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11 program involves general practitioners (63% of patients), hospital physicians (65% of patients),  
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13 nurses (49% of patients), physical therapists (22% of patients). Arterial blood gases (59% of  
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15 patients), sleep study (41% of patients), and nocturnal pulse oximetry (33% of patients), were the  
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17 principal investigations during monitoring program.  
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22 The majority of children with a tracheotomy had uncuffed tube (64,2%). The at home tracheotomy  
23  
24 tube changes were performed by parents (39% of tracheotomised patients), by intensivists (25%),  
25  
26 by nurses (19%), by pulmonologists (10%), or by others (7%). The majority of patients with  
27  
28 tracheotomy (63%) required a tube change every 16-30 days.  
29  
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31  
32 The medical professionals at the reference center were named as the primary provider for home  
33  
34 ventilation management. However, only 48% of children required home health care, mostly those  
35  
36 needing invasive ventilation for  $\geq 12$  hours daily ( $\chi^2_{MH} = 46$ ;  $P < 0.001$ ) (Table 1). Parents were  
37  
38 identified as the first care giver for all the children, and in 171 cases (47%), the sole care giver.  
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40 Other care givers were different professionals (40% of children), family friends (8%), and non  
41  
42 professional volunteers (3%).  
43  
44

45  
46 Pulse oximetry (saturation) was used as monitoring modality. Cardiovascular/respiratory lead  
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48 monitoring was done in 10% of patients, particularly younger children ( $\leq 5$  year age), receiving  
49  
50 non-invasive ventilation, and with neuromuscular disorders.  
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52  
53 Multivariate logistic regression showed that invasive ventilation was likely associated with age of  
54  
55 children - younger patients- (odds ratio [OR] 0.91; 95% confidence interval [CI] 0.84-0.97),  
56  
57 residence in the south of Italy (OR 2.94; CI 1.32-6.55), more years spent in mechanical ventilation  
58  
59 (OR 1.33; CI 1.19-1.47), health care received at home (OR 8.02; CI 3.82-16.84), mechanical  
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3 ventilation  $\geq$  12 hours daily under (OR 11.65; CI 5.92-22.95), or neuromuscular disorder (OR 4.45;  
4  
5 CI 1.13-17.62) or hypoxic (ischemic) encephalopathy (OR 3.62; CI 1.37-9.55).  
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## 10 **DISCUSSION**

11  
12 This is the first report about children receiving long-term home ventilation in Italy. Approximately  
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14 4 per 100.000 children up to 17 years old had chronic mechanical respiratory support at home in  
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16 2007, in good agreement with estimates from other countries.<sup>12,14,21</sup> If the figures are generalized to  
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18 the whole country, a total 404 children in Italy would be found, supporting the reliability of the  
19  
20 present survey involving 362 children. Thus, even if the reported findings do not include all  
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22 possible cases, they do provide the best available information for this group of children. Patients  
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24 were resident in 75 of the 107 Italian provinces (70%), and were mainly discharged from centers in  
25  
26 the North of Italy, which may be attributed to fewer pediatric services in the South, in terms of  
27  
28 quality and quantity.<sup>23</sup> Health inequalities in childhood can lead to distress for the patient and the  
29  
30 family, as well as financial waste. Thus, for this group of patients the burden of care is higher for  
31  
32 southern residents.  
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39 The median length of home ventilation was one year, indicating that hospital discharge of these  
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41 patients is a recent practice in Italy. In line with surveys from other countries<sup>9,12,16,21</sup> neuromuscular  
42  
43 disorders were the most frequent indication for long-term home ventilation, as the established  
44  
45 supportive therapy reduces the morbidity and mortality of these children.<sup>24-28</sup>  
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47

48  
49 Non-invasive support was used in the majority of the investigated population (59%) and was more  
50  
51 prevalent in later infancy and adolescence. This finding is consistent with the literature,<sup>29</sup> which  
52  
53 identifies NIV as the preferred ventilation strategy and recommended IV only if required (i.e.,  
54  
55 inability to swallow, to cooperate, to clear airway secretions). Although, improvements in the  
56  
57 technical features of mechanical ventilators and non-invasive interfaces have made a larger  
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59 proportion of the pediatric population, including very young or small children, suitable for non-  
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invasive mechanical ventilation, the prevalence of non-invasive ventilation in late childhood and adolescence probably reflects the better cooperation by older children.<sup>24,25</sup> However, the paucity of non-invasive interfaces suitable for small children continues to be an important consideration that guides the choice of approach.

Invasive ventilation was significantly related not only to young age, but also to residence in the south, longer time spent under mechanical ventilation, and neuromuscular disorders or hypoxic (ischemic) encephalopathy. Inability to cooperate, to clear airway secretions, and to swallow are more common in neurological disorders and in younger children, and are contraindications to using non-invasive ventilation.<sup>29,30</sup> The survey showed that children affected by neurological disorders provide the vast majority of patients receiving ventilation via tracheotomy. This finding is consistent with the results of Eurovent study.<sup>3</sup> According to criteria proposed by Australasian consensus statement children requiring invasive ventilation spent longer time per day under mechanical ventilation.<sup>31</sup> Regional differences in the type of ventilation reflect the health, social and organizational conditions and the medical staff expertise in the different settings. In fact, in the South of Italy, the vast majority of patients was discharged from pediatric intensive care units.

While NIV is generally accepted to support patients affected by Duchenne muscular dystrophy and SMA type 2,<sup>27,32</sup> the use of NIV for children with SMA type 1 is less clear and respiratory management differs around the world.<sup>33</sup> The benefits and the burdens of both NIV and IV in children affected by SMA type 1 have to be again adequately evaluate.<sup>34</sup> Different views and attitudes have the clinicians recommending/offering the type of ventilation to the parents of children with SMA type 1.<sup>35,36</sup> Findings suggest that in Italy both NIV and IV are used in this population at a similar rate, and further efforts are needs defining appropriate decisions.

One of the most surprising findings of the survey is the relatively high prevalence of children affected by encephalopathy supported with HMV. This finding is consistent with the results of an other Italian study.<sup>10</sup> However, recent surveys documented that in other countries these patients

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2  
3 don't represent one of the most important disorder categories of HMV children.<sup>9,18,21,22</sup> Moreover,  
4  
5 this study showed differences also in the type of interface used to ventilate these children. While in  
6  
7 the United Kingdom NIV is usually preferred,<sup>37</sup> in Italy the majority of children affected by  
8  
9 encephalopathy is invasively ventilated. Both these differences are probably due to a different  
10  
11 ethical approach around the world.  
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14  
15 The main people involved in home ventilation surveys in UK<sup>18</sup> and Massachusetts<sup>22</sup> are pediatric  
16  
17 pulmonologists and general pediatricians, and pediatric intensive care physicians are seldom  
18  
19 involved (11% and 1% of children respectively). In the present survey the majority of patients  
20  
21 (59%) were discharged from pediatric intensive care units or pediatric pulmonary units, suggesting  
22  
23 a different attitude and involvement of Italian general pediatricians in the management of these  
24  
25 patients compared to other countries.  
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29 As expected,<sup>22,38</sup> pulse oximetry was the preferred monitoring modality at home, during non-  
30  
31 invasive and invasive ventilation, particularly for children with neuromuscular disorders. This  
32  
33 approach is in agreement with the regimen for managing acute and chronic neuromuscular  
34  
35 respiratory failure at home described by Bach and colleagues.<sup>24</sup>  
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38  
39 Similarly to French survey<sup>9</sup> follow up was performed during outpatient visits in 60% of the patients.  
40  
41 Records of arterial blood gases, nocturnal pulse oximetry and sleep study were the most common  
42  
43 applied tests during routine follow up. This finding is consistent with the French study.<sup>9</sup>  
44  
45

46  
47 In our study population uncuffed tracheotomy tubes are preferred over cuffed tubes. This finding is  
48  
49 consistent with the literature. In fact, the indication for cuffed tracheotomy tubes are limited (i.e.,  
50  
51 patients with chronic translaryngeal aspiration, patients requiring ventilation with high pressures).<sup>39</sup>  
52

53  
54 There is no consensus regarding the frequency of tracheotomy tube changes. While recommended  
55  
56 tube change frequency ranges from daily to monthly, the most common frequency is weekly.<sup>39</sup>  
57

58  
59 According to the literature, the frequency of tracheotomy tube changes in our population varies  
60  
widely. However, the majority of patients required a tube change every 16-30 days. According to

1  
2  
3 the literature, in most cases the tube change is performed by the caregiver. In fact, before the home  
4 discharge, caregivers should be trained in the replacement of tracheotomy tubes.<sup>39</sup>  
5  
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7  
8 The majority of patients that required home health care need invasive ventilation for more than 12  
9 hours. This finding is consistent with criteria recommended by Australasian consensus statement.<sup>31</sup>  
10  
11 In fact, these children require high level of care and close supervision, which families are unable to  
12 sustain without assistance.  
13  
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16  
17 Heavy workload, loss of personal time, financial, and emotional resources are the main burden of  
18 care related to home care ventilation, falling on parents who, in at least half the cases met all their  
19 child's medical needs themselves. Both the hospital discharge and home care need adequate and  
20 appropriate approaches. Children with chronic ventilation at home need a coordinated  
21 multidisciplinary approach by trained and accountable professionals, parents, and lay people, based  
22 on interactions between different teams from the hospital, community, family, and carers.<sup>18,40-42</sup>  
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32 In our country specific state programs for pediatric home-care are devoted only to children affected  
33 by HIV or cystic fibrosis, while a program of respiratory home care for children with chronic  
34 respiratory failure have not yet been realized. Specific health care plan is already present only in  
35 selected regions.<sup>10,11</sup>  
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41 There are several limitations to the present survey. Although some children may have been  
42 excluded, the data from many different hospital providers give a comprehensive figure of the use of  
43 home mechanical ventilation in pediatric patients in Italy. No attempt was made to collect data  
44 retrospectively to describe the number of children previously ventilated at home or who either died  
45 or were weaned from the ventilator, or switched from invasive to non-invasive ventilation. This will  
46 be one of the topics of future investigations. Families' desires and acceptance of home mechanical  
47 ventilation may be different, and can be influenced by several reasons and factors that have to be  
48 evaluated to guide and support the discharge home. An Italian national ventilator program, and a  
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3 national registry of home-ventilated children could help in monitoring and improving the care of  
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5 these children (and their families) throughout the country.  
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8 In conclusion, the present survey provides, for the first time, reliable estimates of the prevalence of  
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10 children receiving home mechanical ventilation in Italy. The majority of children are noninvasively  
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12 ventilated. Invasive ventilation is significantly related to younger age, regional residence, more  
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14 hours on ventilation and diagnosis (i.e., neurological disorders). Further work is needed to promote  
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16 health program to monitor and improve the wellbeing of these children. However, this is a  
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18 recognized challenge and technical and political commitments are necessary to provide the needed  
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20 resources for patients and their families.  
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**TABLE 1 Children's characteristics and type of ventilation**

Characteristics	Invasive ventilation	Noninvasive ventilation	Total
No. of subjects	149	213	362
Male/female, <i>n</i>	87/62	116/97	203/159 <sup>a1</sup>
Area of residence <sup>b</sup>			
Northern Italy	84	115	199
Central Italy	21	35	56
Southern Italy	44	63	107
Age, median (interquartile range), y	6 (3-9)	11 (5-15)	8 (4-14) <sup>c1</sup>
Age at start of mechanical ventilation			
median (interquartile range), y	1 (0-4)	8 (3-13)	4 (1-11) <sup>c2</sup>
Time on ventilation support			
median (interquartile range), y	3 (1 -5)	1 (0 -2)	1 (0 -4) <sup>c3</sup>
Major diagnostic category <sup>d</sup> , <i>n</i>			
Neuromuscular disorders	66	112	178
Chronic lung and upper respiratory diseases	12	52	64
Hypoxic (ischemic) encephalopathy	31	17	48
Abnormal ventilation control	25	19	44
Spinal cord injury	10	1	11
Chest anomalies	4	12	16
Other	1	0	1
Hours/day spent under mechanical ventilation			
<12 / ≥ 12, <i>n</i>	29/120	178/35	207/155 <sup>a2</sup>
Ventilation monitoring modality, <i>n</i>			
Pulse oximetry (saturation)	136	144	280
Cardiovascular/respiratory leads	8	29	37

End tidal CO <sub>2</sub>	11	0	11
None	3	38	41
Other	4	3	7
Home health care, yes/no, <i>n</i>	121/28	51/162	172/190 <sup>a3</sup>

<sup>a1</sup>:  $\chi^2 = 0.55$ ;  $P > 0.459$ ; <sup>a2</sup>:  $\chi^2 = 148$ ;  $P \ll 0.001$ ; <sup>a3</sup>:  $\chi^2 = 115$ ;  $P \ll 0.001$

<sup>b</sup>:  $\chi^2_{trend} = 0.07$ ;  $P > 0.80$

<sup>c</sup> Mann-Whitney U test: <sup>c1</sup> = -6.147;  $P \ll 0.001$ ; <sup>c2</sup>: = -8.733;  $P \ll 0.001$ ; <sup>c3</sup> = 5.873;  $P \ll 0.001$

<sup>d</sup>:  $F = 5.94$ ;  $P < 0.016$

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For Peer Review

## Long-term Home Ventilation of Children in Italy: a National Survey

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**Funding:** none

**Short title:** Home Mechanical Ventilation for Italian Children

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**Keywords:** Chronic respiratory failure, home respiratory care, long term ventilation, home mechanical ventilation, epidemiology, artificial ventilation, disability.

**Word count:** ~~3019~~

**Number of tables:** 1

**Deleted:** 2962

**Number of figures:** 0

**Conflicts of interest:** none.

For Peer Review

**ABSTRACT**

**Background:** Improved technology, as well as professional and parental awareness, enable many ventilator-dependent children to live at home. However, the profile of this growing population, the quality and adequacy of home care, and patients' needs still require thorough assessment.

**Objectives:** To define the characteristics of Italian children receiving long-term home mechanical ventilation (HMV) in Italy.

**Methods:** A detailed questionnaire was sent to 302 National Health Service hospitals potentially involved in the care of HVM in children (aged < 17 yrs). Information was collected on patient characteristics, type of ventilation, and home respiratory care.

**Results:** A total of 362 HMV children was identified. The prevalence was 4.2 per 100,000 (95% CI: 3.8-4.6), median age was 8 years (interquartile range 4-14), median age at starting mechanical ventilation was 4 years (1-11), and 56% were male. The most frequent diagnostic categories were neuromuscular disorders (49%), lung and upper respiratory tract diseases (18%), hypoxic (ischemic) encephalopathy (13%), and abnormal ventilation control (12%). Medical professionals with nurses (for 62 % of children) and physiotherapists (20 %) participated in the patients' discharge from hospital, though parents were the primary care giver and in 47% of cases, the sole care giver. Invasive ventilation was used in 41% and was significantly related to young age, southern regional residence, longer time spent under mechanical ventilation, neuromuscular disorders, or hypoxic (ischemic) encephalopathy.

**Conclusions:** Care and technical assistance of long-term HMV children need assessment, planning, and resources. A wide variability in pattern of HMV was found throughout Italy. An Italian national ventilation program, as well as a national registry, could be useful in improving the care of these often critically ill children.

## INTRODUCTION

Continuous advances in neonatal and pediatric intensive care are likely to result in more infants, children, and adolescents with critical, chronic conditions, who survive and who are discharged home, but require long-term technological support.<sup>1-3</sup> A growing population of children exists who has chronic respiratory failure due to conditions such as neuromuscular diseases, obstructive sleep apnea, craniofacial abnormalities, or spinal cord injuries, and who require either tracheostomy or mask ventilation.

Pressure to reduce the hospital stay of these children and awareness of the effects of long term hospitalization on a child's development have, during the last decade - especially, in a few countries such as UK, France, and US, - increased the numbers of ventilated children discharged home. However, the availability and practical details of complex respiratory support at home involve a lot of variables such as appropriate home environment, healthcare support, and funding, that cannot always be guaranteed. Findings of the Eurovent study, a wide cross sectional survey including adults as well as children, highlighted how the collection of information about home mechanical ventilation (HMV) could facilitate national planning for HMV. In particular, it could aid the development of HMV services in order to ensure equality of provision and access.<sup>3</sup>

Surveys in Canada,<sup>4,5</sup> Chile,<sup>6</sup> Germany,<sup>7</sup> France,<sup>8,9</sup> Italy,<sup>10,11</sup> Japan,<sup>12</sup> New Zealand,<sup>13</sup> Switzerland,<sup>14</sup> The Netherlands,<sup>15</sup> Turkey,<sup>16</sup> UK,<sup>17-19</sup> and USA<sup>20-22</sup> have defined the size, main features, and needs of the pediatric population requiring home mechanical ventilation. However there are many differences between these observational studies: the setting (single referring center, state, or country), the time period (one year or trend over time), the data collected, and how it was collected. Thus, estimates of the national prevalence of home ventilator-dependent children range from 0.7/100,000 children in 1993 in Japan<sup>12</sup> to 3.4/100,000 in 2000 in Switzerland.<sup>14</sup>

Because information on long-term home ventilation for children in Italy is scant and erratic (no regional or national population-based data exist), this survey was conducted to describe the size and

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2 characteristics of the pediatric Italian population receiving chronic mechanical ventilation at home  
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4 and the profile of home respiratory care.  
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## 7 8 **METHODS**

### 9 10 **Study Population**

11 A questionnaire was sent to all 27 Italian pediatric intensive care and pulmonary units to get data on  
12 children up to 17 years old receiving HMV in January 2007. To identify as many children as  
13 possible the questionnaire was sent also to another 275 hospital-based units (eg. pulmonary  
14 services, and critical care units) of the National Health Service (NHS) that are potentially involved  
15 in the care of the target population. Information referred to children discharged home from hospital  
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### 23 24 **Home respiratory care**

25 HMV was defined as invasive mechanical ventilation (IMV) or non invasive mechanical  
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### 37 38 **Data analysis**

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2 Descriptive statistics were computed for the entire study population and subgroups. Student's *t* test  
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4 was used to compare the type of ventilation and patient's age (continuous variable).  $\chi^2$  tests were  
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6 used to compare the type of ventilation and categorical variables, such as sex and home health care.  
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8 The Mann-Whitney U test was used to compare the type of ventilation in each class of time/day  
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10 spent under mechanical ventilation. ANOVA was used to check the association between the type of  
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12 ventilation and class of diagnosis. Comparisons of the type of ventilation in each region were tested  
13  
14 by  $\chi^2_{\text{trend}}$  analysis. Multivariate logistic regression was used to look at the association between the  
15  
16 type of ventilation and all monitored factors, simultaneously.

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18 Results are presented as frequencies, or median and interquartile range;  $P < 0.05$  was considered  
19  
20 significant.

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22 All data collected on centres and patients were kept strictly confidential. Data on patients were  
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24 anonymous. The San Giovanni Battista-Molinette Hospital, University of Turin, Committee on  
25  
26 Clinical Investigation reviewed and approved the study protocol. Because this was an observational  
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28 study without an intervention and without the presentation of any identifying data, the committee  
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30 ruled that informed consent was not necessary.

## 31 32 33 RESULTS

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35 A total of 362 children were identified by 30 centers (67% in the North of Italy) as receiving home  
36  
37 mechanical ventilation in January 2007, with an estimated prevalence of 4.2 per 100,000  $\leq 17$  year  
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39 old children (95% CI 3.8-4.6). Patients were resident in 75 of the 107 Italian provinces (70%); 200  
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41 (55%) were resident in the North, 55 (15%) in the Center, and 107 (30%) in the South, in good  
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43 agreement with the national distribution of children up to 17 years old. No difference was observed  
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45 between the type of ventilation and the area of residence (Table 1). The majority of patients (59%)  
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47 were discharged from 12 of 27 NHS pediatric intensive care units or pediatric pulmonary units that  
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49 were following mechanically ventilated children at home, and the others came from 18 other

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1 hospital units and wards (i.e., adult intensive care units, adult pulmonary units, pediatric neurology  
2 units, pediatric palliative care units). The median number of children followed by each center was  
3 seven (interquartile range 3-19).

4 The median age was 8 years (interquartile range 4-14) and was significantly different for children  
5 receiving invasive or non-invasive ventilation ( $P \ll 0.001$ ), as well as for the median age at the  
6 start of mechanical ventilation ( $P \ll 0.001$ ) (Table 1). All children received positive pressure  
7 ventilation. The majority of children receives NIV (Table 1), which is delivered mostly by nasal  
8 mask (85% of patients receiving NIV). Non-invasive ventilator support was the most used and  
9 skewed toward late infancy and adolescence.. Overall, 41% of the survey population had ventilation  
10 via tracheotomy with the highest percentage of children affected by neurological disorders. The  
11 median duration of home mechanical ventilation was one year (Table 1).

12 The most frequent diagnostic category was neuromuscular disorders (49%), followed by lung and  
13 upper respiratory diseases (18%), hypoxic (ischemic) encephalopathy (13%), and abnormal  
14 ventilation control (12%) (Table 1). The most represented neuromuscular disorders were spinal  
15 muscular atrophy (SMA) (78 patients), Duchenne muscular dystrophy (31 patients) and Congenital  
16 muscular dystrophy (29 patients). In particular, 25 children were affected by SMA type 1 (16  
17 receiving NIV, 64%) and 53 children by SMA type 2 (30 receiving NIV, 57%).

18 There were wide difference between the type of ventilation and diagnoses (  $F = 5.94$ ,  $P < 0.016$ ).

19 Time per day spent under mechanical ventilation was longer for children requiring invasive  
20 ventilation ( $\chi^2 = 148$ ;  $P \ll 0.001$ ) (Table 1), particularly for those with neuromuscular disorders ( $\chi^2$   
21 = 73;  $P \ll 0.001$ ), hypoxic (ischemic) encephalopathy ( $\chi^2 = 37$ ;  $P \ll 0.001$ ), or chronic lung and  
22 upper respiratory diseases ( $\chi^2 = 28$ ;  $P \ll 0.001$ ).

23 Nurses (for 62 % of children), physiotherapists (20 %), social workers (7 %), psychologists (7 %),  
24 pediatricians (5%), and other professionals (4%) participated in the home discharge process with  
25 hospital physicians. All children were involved in a patient ventilator training program (94% during  
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2 inpatient visits; 6% during outpatient visits) and in a regular monitoring program. At home visits  
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4 were planned in 60% of patients (36% of patients only during planned hospitalization, 30% only  
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6 during outpatient visits, 29% with both modalities, 5 and % only at home). The mean number of  
7  
8 hospital visits was 2.8/year, while the mean number of at home visits was 27/year. The at home visit  
9  
10 program involves general practitioners (63% of patients), hospital physicians (65% of patients),  
11  
12 nurses (49% of patients), physical therapists (22% of patients). Arterial blood gases (59% of  
13  
14 patients), sleep study (41% of patients), and nocturnal pulse oximetry (33% of patients), were the  
15  
16 principal investigations during monitoring program.

17  
18 The majority of children with a tracheotomy had uncuffed tube (64,2%). The at home tracheotomy  
19  
20 tube changes were performed by parents (39% of tracheotomised patients), by intensivists (25%),  
21  
22 by nurses (19%), by pulmonologists (10%), or by others (7%). The majority of patients with  
23  
24 tracheotomy (63%) required a tube change every 16-30 days.

25  
26 The medical professionals at the reference center were named as the primary provider for home  
27  
28 ventilation management. However, only 48% of children required home health care, mostly those  
29  
30 needing invasive ventilation for  $\geq 12$  hours daily ( $\chi^2_{MH} = 46$ ;  $P \ll 0.001$ ) (Table 1). Parents were  
31  
32 identified as the first care giver for all the children, and in 171 cases (47%), the sole care giver.  
33  
34 Other care givers were different professionals (40% of children), family friends (8%), and non  
35  
36 professional volunteers (3%).

37  
38 Pulse oximetry (saturation) was used as monitoring modality. Cardiovascular/respiratory lead  
39  
40 monitoring was done in 10% of patients, particularly younger children ( $\leq 5$  year age), receiving  
41  
42 non-invasive ventilation, and with neuromuscular disorders.

43  
44 Multivariate logistic regression showed that invasive ventilation was likely associated with age of  
45  
46 children - younger patients- (odds ratio [OR] 0.91; 95% confidence interval [CI] 0.84-0.97),  
47  
48 residence in the south of Italy (OR 2.94; CI 1.32-6.55), more years spent in mechanical ventilation  
49  
50 (OR 1.33; CI 1.19-1.47), health care received at home (OR 8.02; CI 3.82-16.84), mechanical

8

1  
2 ventilation  $\geq$  12 hours daily under (OR 11.65; CI 5.92-22.95), or neuromuscular disorder (OR 4.45;  
3  
4 CI 1.13-17.62) or hypoxic (ischemic) encephalopathy (OR 3.62; CI 1.37-9.55).  
5  
6

## 7 8 **DISCUSSION**

9  
10 This is the first report about children receiving long-term home ventilation in Italy. Approximately  
11  
12 4 per 100.000 children up to 17 years old had chronic mechanical respiratory support at home in  
13  
14 2007, in good agreement with estimates from other countries.<sup>12,14,21</sup> If the figures are generalized to  
15  
16 the whole country, a total 404 children in Italy would be found, supporting the reliability of the  
17  
18 present survey involving 362 children. Thus, even if the reported findings do not include all  
19  
20 possible cases, they do provide the best available information for this group of children. Patients  
21  
22 were resident in 75 of the 107 Italian provinces (70%), and were mainly discharged from centers in  
23  
24 the North of Italy, which may be attributed to fewer pediatric services in the South, in terms of  
25  
26 quality and quantity.<sup>23</sup> Health inequalities in childhood can lead to distress for the patient and the  
27  
28 family, as well as financial waste. Thus, for this group of patients the burden of care is higher for  
29  
30 southern residents.

31  
32 The median length of home ventilation was one year, indicating that hospital discharge of these  
33  
34 patients is a recent practice in Italy. In line with surveys from other countries<sup>9,12,16,21</sup> neuromuscular  
35  
36 disorders were the most frequent indication for long-term home ventilation, as the established  
37  
38 supportive therapy reduces the morbidity and mortality of these children.<sup>24-28</sup>

39  
40 Non-invasive support was used in the majority of the investigated population (59%) and was more  
41  
42 prevalent in later infancy and adolescence. This finding is consistent with the literature,<sup>29</sup> which  
43  
44 identifies NIV as the preferred ventilation strategy and recommended IV only if required (i.e.,  
45  
46 inability to swallow, to cooperate, to clear airway secretions). Although, improvements in the  
47  
48 technical features of mechanical ventilators and non-invasive interfaces have made a larger  
49  
50 proportion of the pediatric population, including very young or small children, suitable for non-

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invasive mechanical ventilation, the prevalence of non-invasive ventilation in late childhood and adolescence probably reflects the better cooperation by older children.<sup>24,25</sup> However, the paucity of non-invasive interfaces suitable for small children continues to be an important consideration that guides the choice of approach.

Invasive ventilation was significantly related not only to young age, but also to residence in the south, longer time spent under mechanical ventilation, and neuromuscular disorders or hypoxic (ischemic) encephalopathy. Inability to cooperate, to clear airway secretions, and to swallow are more common in neurological disorders and in younger children, and are contraindications to using non-invasive ventilation.<sup>29,30</sup> The survey showed that children affected by neurological disorders provide the vast majority of patients receiving ventilation via tracheotomy. This finding is consistent with the results of Eurovent study.<sup>3</sup> According to criteria proposed by Australasian consensus statement children requiring invasive ventilation spent longer time per day under mechanical ventilation.<sup>31</sup> Regional differences in the type of ventilation reflect the health, social and organizational conditions and the medical staff expertise in the different settings. In fact, in the South of Italy, the vast majority of patients was discharged from pediatric intensive care units.

While NIV is generally accepted to support patients affected by Duchenne muscular dystrophy and SMA type 2,<sup>27,32</sup> the use of NIV for children with SMA type 1 is less clear and respiratory management differs around the world.<sup>33</sup> The benefits and the burdens of both NIV and IV in children affected by SMA type 1 have to be again adequately evaluate.<sup>34</sup> Different views and attitudes have the clinicians recommending/offering the type of ventilation to the parents of children with SMA type 1.<sup>35,36</sup> Findings suggest that in Italy both NIV and IV are used in this population at a similar rate, and further efforts are needs defining appropriate decisions.

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One of the most surprising findings of the survey is the relatively high prevalence of children affected by encephalopathy supported with HMV. This finding is consistent with the results of an other Italian study.<sup>10</sup> However, recent surveys documented that in other countries these patients

10

1 don't represent one of the most important disorder categories of HMV children.<sup>9,18,21,22</sup> Moreover,  
2  
3 this study showed differences also in the type of interface used to ventilate these children. While in  
4  
5 the United Kingdom NIV is usually preferred,<sup>37</sup> in Italy the majority of children affected by  
6  
7 encephalopathy is invasively ventilated. Both these differences are probably due to a different  
8  
9 ethical approach around the world.

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11 The main people involved in home ventilation surveys in UK<sup>18</sup> and Massachusetts<sup>22</sup> are pediatric  
12  
13 pulmonologists and general pediatricians, and pediatric intensive care physicians are seldom  
14  
15 involved (11% and 1% of children respectively). In the present survey the majority of patients  
16  
17 (59%) were discharged from pediatric intensive care units or pediatric pulmonary units, suggesting  
18  
19 a different attitude and involvement of Italian general pediatricians in the management of these  
20  
21 patients compared to other countries.

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23 As expected,<sup>22,38</sup> pulse oximetry was the preferred monitoring modality at home, during non-  
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25 invasive and invasive ventilation, particularly for children with neuromuscular disorders. This  
26  
27 approach is in agreement with the regimen for managing acute and chronic neuromuscular  
28  
29 respiratory failure at home described by Bach and colleagues.<sup>24</sup>

31 Similarly to French survey<sup>9</sup> follow up was performed during outpatient visits in 60% of the patients.  
32  
33 Records of arterial blood gases, nocturnal pulse oximetry and sleep study were the most common  
34  
35 applied tests during routine follow up. This finding is consistent with the French study.<sup>9</sup>

36 In our study population uncuffed tracheotomy tubes are preferred over cuffed tubes. This finding is  
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38 consistent with the literature. In fact, the indication for cuffed tracheotomy tubes are limited (i.e.,  
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40 patients with chronic translaryngeal aspiration, patients requiring ventilation with high pressures).<sup>39</sup>

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42 There is no consensus regarding the frequency of tracheotomy tube changes. While recommended  
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44 tube change frequency ranges from daily to monthly, the most common frequency is weekly.<sup>39</sup>

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46 According to the literature, the frequency of tracheotomy tube changes in our population varies  
47  
48 widely. However, the majority of patients required a tube change every 16-30 days. According to

1  
2 the literature, in most cases the tube change is performed by the caregiver. In fact, before the home  
3 discharge, caregivers should be trained in the replacement of tracheotomy tubes.<sup>39</sup>

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6 The majority of patients that required home health care need invasive ventilation for more than 12  
7 hours. This finding is consistent with criteria recommended by Australasian consensus statement.<sup>31</sup>

8  
9 In fact, these children require high level of care and close supervision, which families are unable to  
10 sustain without assistance.

11  
12 Heavy workload, loss of personal time, financial, and emotional resources are the main burden of  
13 care related to home care ventilation, falling on parents who, in at least half the cases met all their  
14 child's medical needs themselves. Both the hospital discharge and home care need adequate and  
15 appropriate approaches. Children with chronic ventilation at home need a coordinated  
16 multidisciplinary approach by trained and accountable professionals, parents, and lay people, based  
17 on interactions between different teams from the hospital, community, family, and carers.<sup>18,40-42</sup>

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18  
19 In our country specific state programs for pediatric home-care are devoted only to children affected  
20 by HIV or cystic fibrosis, while a program of respiratory home care for children with chronic  
21 respiratory failure have not yet been realized. Specific health care plan is already present only in  
22 selected regions.<sup>10,11</sup>

23  
24  
25 There are several limitations to the present survey. Although some children may have been  
26 excluded, the data from many different hospital providers give a comprehensive figure of the use of  
27 home mechanical ventilation in pediatric patients in Italy. No attempt was made to collect data  
28 retrospectively to describe the number of children previously ventilated at home or who either died  
29 or were weaned from the ventilator, or switched from invasive to non-invasive ventilation. This will  
30 be one of the topics of future investigations. Families' desires and acceptance of home mechanical  
31 ventilation may be different, and can be influenced by several reasons and factors that have to be  
32 evaluated to guide and support the discharge home. An Italian national ventilator program, and a

1  
2 national registry of home-ventilated children could help in monitoring and improving the care of  
3  
4 these children (and their families) throughout the country.

5  
6 In conclusion, the present survey provides, for the first time, reliable estimates of the prevalence of  
7  
8 children receiving home mechanical ventilation in Italy. The majority of children are noninvasively  
9  
10 ventilated. Invasive ventilation is significantly related to younger age, regional residence, more  
11  
12 hours on ventilation and diagnosis (i.e., neurological disorders). Further work is needed to promote  
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14 health program to monitor and improve the wellbeing of these children. However, this is a  
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16 recognized challenge and technical and political commitments are necessary to provide the needed  
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18 resources for patients and their families.

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TABLE 1 Children's characteristics and type of ventilation

Characteristics	Invasive ventilation	Noninvasive ventilation	Total
No. of subjects	149	213	362
Male/female, <i>n</i>	87/62	116/97	203/159 <sup>a1</sup>
Area of residence <sup>b</sup>			
Northern Italy	84	115	199
Central Italy	21	35	56
Southern Italy	44	63	107
Age, median (interquartile range), <i>y</i>	6 (3-9)	11 (5-15)	8 (4-14) <sup>c1</sup>
Age at start of mechanical ventilation			
median (interquartile range), <i>y</i>	1 (0-4)	8 (3-13)	4 (1-11) <sup>c2</sup>
Time on ventilation support			
median (interquartile range), <i>y</i>	3 (1 -5)	1 (0 -2)	1 (0 -4) <sup>c3</sup>
Major diagnostic category <sup>d</sup> , <i>n</i>			
Neuromuscular disorders	66	112	178
Chronic lung and upper respiratory diseases	12	52	64
Hypoxic (ischemic) encephalopathy	31	17	48
Abnormal ventilation control	25	19	44
Spinal cord injury	10	1	11
Chest anomalies	4	12	16
Other	1	0	1
Hours/day spent under mechanical ventilation			
<12 / ≥ 12, <i>n</i>	29/120	178/35	207/155 <sup>a2</sup>
<u>Ventilation monitoring modality, <i>n</i></u>			
<u>Pulse oximetry (saturation)</u>	<u>136</u>	<u>144</u>	<u>280</u>
<u>Cardiovascular/respiratory leads</u>	<u>8</u>	<u>29</u>	<u>37</u>

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<u>End tidal CO<sub>2</sub></u>	<u>11</u>	<u>0</u>	<u>11</u>
<u>None</u>	<u>3</u>	<u>38</u>	<u>41</u>
<u>Other</u>	<u>4</u>	<u>3</u>	<u>7</u>
Home health care, yes/no, <i>n</i>	121/28	51/162	172/190 <sup>a3</sup>

<sup>a1</sup>:  $\chi^2=0.55$ ;  $P > 0.459$ ; <sup>a2</sup>:  $\chi^2 = 148$ ;  $P \ll 0.001$ ; <sup>a3</sup>:  $\chi^2 = 115$ ;  $P \ll 0.001$

<sup>b</sup>:  $\chi^2_{trend} = 0.07$ ;  $P > 0.80$

<sup>c</sup> Mann-Whitney U test: <sup>c1</sup> = -6.147;  $P \ll 0.001$ ; <sup>c2</sup>: = -8.733;  $P \ll 0.001$ ; <sup>c3</sup> = 5.873;  $P \ll 0.001$

<sup>d</sup>:  $F = 5.94$ ;  $P < 0.016$

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Deleted: TABLE 2 Ventilation monitoring modality according to the type of ventilation used¶  
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Modality ... [1]

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For Peer Review

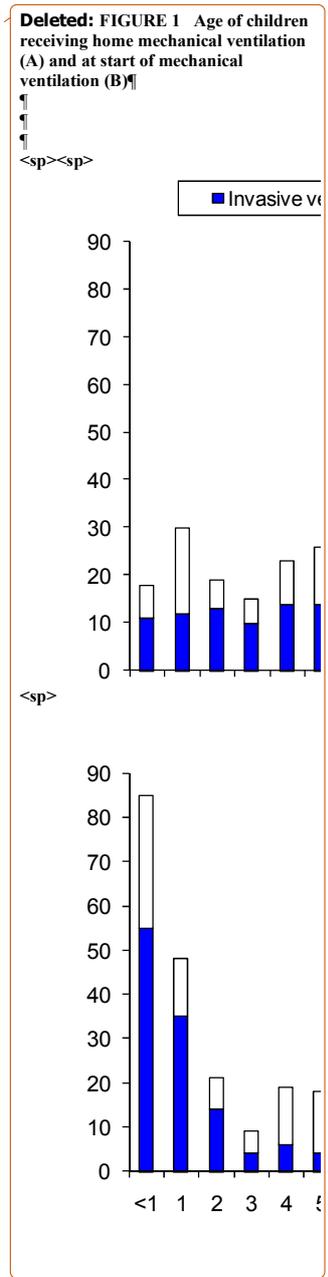


TABLE 2 Ventilation monitoring modality according to the type of ventilation used

Modality	Invasive ventilation <i>n</i> (%)	Noninvasive ventilation <i>n</i> (%)	Total <i>n</i> (%)
Pulse oximetry (saturation)	136 (84)	144 (67)	280 (74)
Cardiovascular/respiratory leads	8 (5)	29 (14)	37 (10)
End tidal CO <sub>2</sub>	11(7)	0	11 (3)
None	3 (2)	38 (18)	41 (11)
Other	4 (2)	3 (1)	7 (2)

The figures do not add up to 100% because some patients used more than one monitoring modality.