Impact of the Regional Pediatric Palliative Care Network on the Care of Children on Long-Term Ventilation: Could the Availability of a Residential Solution into the Network Reduce the Duration of Intensive Care Unit Staying for These Patients?

Francesca Rusalen¹ Caterina Agosto¹ Luca Brugnaro² Franca Benini¹

¹ Pediatric Palliative Care and Pain Service, Pediatric Hospice, Department of Woman and Child's Health, University of Padua, Padua, Italy

² Department of Education and Training, University Hospital Padua, Padua, Italy

Address for correspondence Francesca Rusalen, MD, Pediatric Palliative Care and Pain Service, Pediatric Hospice, Department of Woman and Child's Health, University of Padua, Via Giustiniani 3, 35127 Padua, Italy (e-mail: rusalen.francesca@libero.it).

J Pediatr Intensive Care 2018;7:75–80.

Abstract

The prevalence of children on long-term ventilation (LTV) at home has increased in many countries. In Italy, there are 4.3/100,000 population younger than 18 years. Pediatric palliative care (PPC) network provides high-level care for these patients. In December 2003, in the northeast region of Italy, the regional authority promoted and developed a regional network for PPC, a regional network dedicated to the management of pediatric patients with life-limiting and life-threatening diseases. Characterization of LTV children population and description of care offered to them by a regional PPC network, based on the experience of the Veneto region were collected in a regional database. The regional database and evaluation of families' satisfaction, by means of a questionnaire, were longitudinally analyzed. We studied 56 children on LTV. The main involved diseases were neuromuscular diseases and myopathy. All patients had major comorbidities. Mean age was 4.5 years. The median age of starting ventilation was 3.9 years. The initial type of ventilation was invasive mechanical ventilation in 31 patients and noninvasive ventilation in 25 of them. The overall average frequency of hospitalizations for acute episodes was 0.7 admissions per year and the median duration of staying was 6 days. The median duration of staying in the intensive care unit (ICU) was significantly reduced after the opening of a residential solution (the pediatric hospice) (5 vs. 39 days). PPC network can offer global care to children on LTV. The availability of a residential structure into the PPC network seems to reduce the number and duration of hospital stays, especially in ICU, of this population, with likely cost savings. Further studies are necessary to confirm this hypothesis.

Keywords

- long-term ventilation
- mechanical ventilation
- Pediatric Palliative Care
- life-limiting disease
- intensive care

received
January 8, 2017
accepted after revision
July 8, 2017
published online
August 11, 2017

Copyright © 2018 by Georg Thieme
Verlag KG, Stuttgart · New York

DOI <https://doi.org/10.1055/s-0037-1605369>.
ISSN 2146-4618.

Introduction

In the last 20 years, there has been a gradual increase in the prevalence of pediatric patients on long-term ventilation (LTV) at home,^{1,2} both invasive mechanical ventilation (IMV) and noninvasive ventilation (NIV).^{2,3} In Italy, the prevalence is 4.3/100,000 population of children younger than 18 years, and 70% of them are under the care of pediatric specialists, mainly pediatric pulmonologists or anesthesiologists.⁴ The needs of this population are complex and call for solutions by multidisciplinary teams.^{5,6} A pediatric palliative care (PPC) dedicated network⁷ can facilitate the global management of these patients, and it might represent a reference care model for this population. A PPC network, providing assistance at home, as well as residential solution (pediatric hospice), and coordinating the health providers, can respond to the complex and different needs of these patients, offering a continuous and central reference service for both health care providers and children and families.

Patients on LTV at home account for a significant proportion of the population receiving PPC, estimated in the literature in the range of 50 to 90%.^{1,8,9} The literature proposes different models of care for this population. In Italy, there are no national registers or shared programs; therefore, there are important regional differences about availability and accessibility of services of care around the country.⁴

In December 2003, the Veneto Regional Authority was the first in Italy to promote and develop the regional network for PPC, a regional network dedicated to the management of pediatric patients with life-limiting and life-threatening diseases. The assumptions for the creation of the PPC network were the National Italian Law 38/2010, which set PPC as an inalienable right, and the regional deliberation on the modalities of concrete realization of the network. In the network, only one hospice facility was instituted, in reason of both the high required expertise of dedicated team and the little population of children involved. The location of this facility has been chosen centrally in the region close to the Regional Central Hospital. The regional PPC network includes the Regional PPC Center, the Regional Central Hospital, the nearest local hospitals, the local district health services, and the local pediatricians, which create a network of care around these children and their families. The Veneto Regional Center for PPC and Pain Treatment (PPC Center) has a function of organizing, coordinating, and supporting the activities and interventions of the regional network of PPC for the whole Veneto region (a northern Italian region with 5 million inhabitants in an area of 18,264 km²). Since 2009, the pediatric hospice, a residential solution, has been included into the PPC network, as part of the PPC Center.

The PPC Center guarantees 24-hour-day assistance to all these patients in the Veneto region, coordinating the activities of the network and offering a multidisciplinary team that responds competently to the different needs (clinical, psychological, organizational, social, and spiritual) of these children and their families, both at home and at school, as well as both in the hospital and in the pediatric hospice. The

PPC network takes care of these children, both during acute events and follow-up (FU), choosing the most appropriate setting for the child's care in the network thanks to the PPC Center's coordination.

The PPC Center ensures regularly FU to these patients on the respiratory conditions and the mechanical ventilation, as well as on the others comorbidities, as well as also on the psychological, social, and spiritual needs, with a global highly specialized care, offering dedicated solutions. This can help prevent acute events, or anyway to early treat these acute events. In addition, the residential solution offers an alternative familiar setting where the child can be treated as soon as the critical conditions are resolved, or when the palliative path is required also during these acute events (such as for patients with do-not-resuscitate order).

Aims

The primary aim of this study was to identify the demographic and clinical characteristics of this population and to describe the care offered by the regional PPC network. The secondary aim was to assess the impact of the Veneto PPC network on the care of the children on LTV at home.

Materials and Methods

Study Design

This is a longitudinal descriptive analysis.

Population

Patients on domiciliary LTV in the care of the PPC Center from January 1, 2004, to December 31, 2013 (including patients who were discharged or died during this time) were included. Two different periods were considered: the first period (2004–2008) consisted in the first 5 years of the network activity, when any residential solution was not yet available, and the second period (2009–2013), when the residential solution (the pediatric hospice) was included into the network.

Inclusion Criteria

Children needing IMV or NIV support, for at least 3 months, in stable clinical conditions, managed at home were included.

Variables Considered

Gender and age were the demographic characteristics in this study. Clinical characteristics were principal diagnosis and comorbidities, age of starting ventilation, type of ventilation (IMV vs. NIV), respiratory and motor physical therapy. Clinical outcomes were frequency and duration of hospitalization for acute episodes, frequency and duration of staying in intensive care unit (ICU) during acute events, and frequency and duration of hospitalization for follow-up (FU). We have also compared the frequency and the duration of hospitalization, particularly the frequency and duration of staying at the ICU, during acute episodes, before and after the pediatric hospice opening. Lastly, the population survival and the place of death were also considered.

Tools

We conducted a descriptive longitudinal analysis based on a regional database of the variables considered. For the purpose, a suitable regional database has been created, which has been filled in with the data collected from regular clinical records, both printed and computer-based records, obtained during both the acute events and the FU hospitalization, as well as from the computerized records of the domiciliary activities conducted by the PPC Center. The data were collected by the health care staff of the PPC Center. In addition, we have utilized an anonymous questionnaires for inquiring families' satisfaction.

Statistical Analysis

Primary Outcome

Description of main characteristics of the study cohort and care offered by the Regional PPC Network was evaluated.

Secondary Outcomes

Frequency and duration of hospitalizations for children on LTV, cared by the PPC network, and comparison between the early period (January 1, 2004–December 31, 2008) and the latest period (January 1, 2009–December 31, 2013), after the pediatric hospice opening were evaluated.

Descriptive Analysis

Absolute and relative frequencies for the qualitative variables, mean $\pm$ standard deviation for the normally distributed quantitative variables, and principal quartiles (median, first, and third quartiles) for quantitative variables violating the assumption of a normal distribution (Shapiro–Wilk's test) were analyzed.

Group Comparisons

In this study we here used the *t*-test for comparison of normally distributed quantitative variables, the Wilcoxon–Mann–Whitney's test for quantitative variables found not normally distributed, the log-rank test for the survival analysis, and the chi-square test on the hypothesis of independence for the qualitative variables. A $p < 0.05$ was considered statistically significant. The R software version 3.0.1 and Microsoft Excel 2007 were used to perform the statistical analysis.

Results

Our sample resulted 56 patients with median age of 3.2 years (± 4.9 years) (range: 0–18 years), and 55% of them were males. The main involved diseases were neuromuscular diseases (NMDs) and myopathy (for other involved diseases, see ►Table 1). All the considered children presented comorbidities (see ►Table 1). These patients were addressed to the PPC network by pediatric anesthesiologists (43%), pediatric specialists (23%), pediatricians at hospital (13%), local pediatricians (11%), neonatologists (4%), or other health care providers (5%).

The median age of starting ventilation was 3.9 years; 70% of them have started the ventilation during an acute event and 30% in an elective situation. The initial type of ventilation was IMV in 31 patients (55%) and NIV in 25 patients (45%). The starting of ventilation was associated with respiratory physical therapy in 29 patients (52%). In addition, physical therapy was routinely performed on 40 patients (71%).

At the end of the period considered for the study, 38 patients (64.3%) were still on the care of the PPC Center; 10 patients had died (17.9%); and 8 had discharged (14.3%). As on December 31, 2013, the patients on LTV at home accounted for 50% of all the children managed by the center (38/76 patients).

Table 1 Characteristics of the sample

Total number 56 pts		
Gender	Male 31 pts	Female 25 pts
Median age	3.2 y (± 4.9)	Range: 0–18 y
Principal diagnosis	17 neuromuscular diseases 11 myopathy 7 congenital central hypoventilation syndrome 7 spinal nerve disorders 4 complex cardiopathies 3 metabolic disorders 2 oncohematological diseases 5 other conditions (2 sequelae of extreme prematurity, 2 trisomies, and 1 congenital hepatic arteriovenous malformation)	
Comorbidities (100% of pts)	96% neuromotor 71% osteoarticular 70% gastroenterological 36% cardiological 32% neurocognitive 23% sensory (sight and/or hearing) 20% nephrological	

Abbreviation: pts, patients.

We compared the frequency and duration of hospitalization, both for acute (including at the ICU) and FU, considering the two different periods, before and after the opening of the pediatric hospice. There was no significant difference, regarding age, gender, and main disease, between the populations considered in these two periods.

The overall average frequency of hospitalization for acute episodes was 0.7 admissions per year and the median duration of staying was 6 days (range: 1–103). There was no significant difference after the opening of the pediatric hospice (frequency: pre—0.6 admissions per year and post—0.7 admissions per year, and duration: pre—7 days and post—6 days).

The mean number of ICU admission was 0.075 per patient per year, with a linear correlation, statistically significant ($p = 0.008$) between number of patients admitted at the ICU per year and number of patients managed by the PPC Center per year. There was no significant difference after the opening of the pediatric hospice.

The median duration of staying in the ICU was 7 days (range: 2–79), with a difference after the pediatric hospice's opening: pre—39 days (range: 7–96) and post—5 days (range: 2–79) ($p = 0.07$) (see ►Fig. 1).

The overall average frequency of hospitalization for FU was 2.3 admissions per year. The median duration of hospitalization for FU was 2 days (range: 1–34), in both the considered periods. The overall average frequency of hospitalization, considering both acute and FU hospitalizations, was three admissions per year. The median of total time spent in hospital was 9 days per year (range: 2–96), considering both acute and FU hospitalizations.

The global survival rate of our sample was 61% at 10 years. The survival data have highlighted a high variability in relation to the main disease: there has been a greater survival for NMDs, myopathy, congenital central hypoventilation syndrome, and neuropathy/spinal diseases (78% over 11

years) ($p = 0.041$). In 14.3% of cases (eight patients), the patient was discharged by the PPC Center care.

Concerning the end-of-life care, 10 children died: 7 of them outside the hospital (3 in the pediatric hospice and 4 at home) and 3 in hospital (2 children in the ICU and 1 in emergency room). All these patients received palliative care during terminal phase of the disease.

The families' satisfaction about services offered by PPC Center was high (84% excellent, 9.5% satisfactory, and 6.5% sufficient). Families have recognized the usefulness of PPC Center in relation to guarantee continuity of care (28%), ensure a shared care plan (22%), represent the common reference for the network (20%), and offer a centralized coordination (14%), for guaranteeing a psychological support (13%).

Discussion

The severe clinical conditions of these children cause repeated admissions at hospital for both diagnostics and treatments, frequently for acute and life-threatening exacerbations, demanding intervention of different specialist pediatricians. Moreover, all these patients present severe comorbidities, as generally also reported in the literature,^{1,3,10} apart from a lower frequency of comorbidities in reports from the United Kingdom (67%).¹¹ The complexity of these patients' management requires highly specialized expertise and it is also essential to ensure an ongoing global care for this population, either at the hospital or at home. For that, these patients need for ad hoc care services with integrated multidisciplinary teams and call for solutions from different institutional agencies. A network model, with a constant reference to a high-level specialized center, might offer to these patients and their families a global care and more safe life at home.

Concerning the hospitalizations, in our sample, the number of hospital admissions for acute events was considerably lower than in a similar study conducted on children eligible for PPC in 2002¹²: before the PPC network was established, for these children, the median number of hospitalizations was 9.7 admissions per year in 2002, as opposed to 3 admissions per year (FU and acute episodes combined, in our study) after the regional PPC network's creation. Therefore, the PPC regional network might enable a reduction in frequency and duration of our patients' hospital staying,^{13–15} with a likely consequent positive effect on the public health costs.¹³ In addition, also the median of total time spent in hospital per year was much higher before the PPC network's creation, amounting to 150 days per year, whereas after that, the median was less than 10 days. The availability of a residential solution into the PPC network could also markedly reduce the duration of ICU staying for these children, with a great difference after the pediatric hospice opening.

This reduction could be partly an effect of the specific expertise developed at the pediatric hospice for managing these complex patients and partly an effect of the careful clinical monitoring which reduces morbidity and complications in these complex patients. The residential solution

Fig. 1 Duration of staying in ICU. Comparison of the median period of ICU staying in relation to the availability of a residential structure in the PPC network: the graph illustrates the decreasing of the duration of staying at the ICU after the pediatric hospice opening. ICU, intensive care unit; PPC, pediatric palliative care.

could represent an optimal alternative to ICU staying for these children during acute episodes. The availability of a residential solution in the PPC network not only guarantees the continuity of care ensuring respect of the shared advance care planing but also offers adequate specific competences for critical acute exacerbations in these children, in a more familiar environmental, also allowing parents to be closer to the child. The possibility to stay at the pediatric hospice during critical acute exacerbations was offered for children with “do-not-resuscitate” order. In addition, for children without this order, the residential solution has represented a possibility of an early discharge from the ICU, ensuring to the child to be care in a dedicated structure with wide competences related with complex patients on LTV.

Further studies are absolutely necessary to verify our hypothesis, particularly because the result of the decreasing duration of ICU staying is not statistically significant, probably because of the limited sample size, due to the rare nature of the diseases involved, but also because of the worsening clinical condition of some patients, consequent to the natural history of these diseases.

To improve the quality of further studies, it might be useful to realize a multicenter study that likely could ensure a more adequate sample size, but at the same time, this kind of study could be greatly limited by the absence of a national Italian register and shared programs, with important regional differences about availability and accessibility of services of care around the Italian country.

The mortality rate in our sample was considerably lower than the figure reported elsewhere for pediatric patients receiving PPC (30% at 1 year),⁸ but this might be also probably because of the characteristics and diagnosis of the patients enrolled. In any case, our study has also confirmed, as reported in the literature, that the overall survival for children with incurable disease can be better, if they receive PPC early.

The place of death that we can consider as an indirect indicator of quality of care has been almost always outside the hospital. Indeed, home and pediatric hospice are widely considered as the better places of end-of-life care for children with incurable disease.^{14,16} In our sample, the percentage of deaths outside the hospital was very high.¹⁰ In addition, it has already been widely recognized that the home, at first, and the hospice, at second, are considered as the better places of care for patients with life-limiting and life-threatening diseases.

Our study presents several limitations: the most important is analysis of a regional reality, so not giving necessarily the possibility to extrapolate conclusion on nationwide. In addition, it is a descriptive study, allowing only to make hypothesis and not clearly define nationwide conclusions. Moreover, the limited sample size has important statistical implications.

Conclusion

A PPC network can help improve the care of children on domiciliary LTV, offering both a constant clinical monitoring

and adequate care during acute events or emergency situations. By that, the PPC network seems to reduce the median time spent at the hospital, so also the number and duration of hospitalization for acute events, optimizing the use of the resources available in the territory. Especially, the presence of a residential solution (such as a pediatric hospice) in the network seems to contribute to reduce the duration of ICU staying for these patients. Future prospective studies are needed to verify this hypothesis and more to also precisely evaluate probable cost savings.

Note

All procedures performed in this study, involving human participants, have been in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The study has obtained the approval of the Ethics Committee. Informed consent has been obtained from all individual participants included in the study. The study has been performed at the Pediatric Pain and Palliative Care Service, Department of Pediatrics, University of Padua, Via Giustiniani 3, 35127 Padua, Italy.

Authors' Contribution

Francesca Rusalen, MD—literature search, data collection, study design, analysis of data, article preparation, and review of the article.

Caterina Agosto, MD—literature search, study design, and review of the article.

Luca Brugnaro, PhD—analysis of data.

Franca Benini, MD—study design and review of the article.

Funding

None.

Conflict of Interest

None.

References

- Goodwin S, Smith H, Langton Hewer S, et al. Increasing prevalence of domiciliary ventilation: changes in service demand and provision in the South West of the UK. *Eur J Pediatr* 2011;170(09):1187–1192
- McDougall CM, Adderley RJ, Wensley DF, Seear MD. Long-term ventilation in children: longitudinal trends and outcomes. *Arch Dis Child* 2013;98(09):660–665
- Com G, Kuo DZ, Bauer ML, et al. Outcomes of children treated with tracheostomy and positive-pressure ventilation at home. *Clin Pediatr (Phila)* 2013;52(01):54–61
- Racca F, Berta G, Sequi M, et al; LTV Pediatric Italian Network. Long-term home ventilation of children in Italy: a national survey. *Pediatr Pulmonol* 2011;46(06):566–572
- Graham RJ. Long-term ventilation: charting a pathway home. *Arch Dis Child* 2013;98(09):653
- Jardine E, Wallis C; UK Working Party on Paediatric Long Term Ventilation. Core guidelines for the discharge home of the child on long-term assisted ventilation in the United Kingdom. *Thorax* 1998;53(09):762–767
- Ferrante A, Po' C, Del Favero E, et al. Does a pediatric palliative care network respond to the needs of families? Efficacy of a pediatric palliative network. *Pediatr Anesth Crit Care J* 2013;1:15–26

- 8 Feudtner C, Kang TI, Hexem KR, et al. Pediatric palliative care patients: a prospective multicenter cohort study. *Pediatrics* 2011; 127(06):1094–1101
- 9 Peterson-Carmichael SL, Cheifetz IM. The chronically critically ill patient: pediatric considerations. *Respir Care* 2012;57(06): 993–1002, discussion 1002–10038
- 10 Amin R, Sayal P, Syed F, Chaves A, Moraes TJ, MacLusky I. Pediatric long-term home mechanical ventilation: twenty years of follow-up from one Canadian center. *Pediatr Pulmonol* 2014;49(08):816–824
- 11 Wallis C, Paton JY, Beaton S, Jardine E. Children on long-term ventilatory support: 10 years of progress. *Arch Dis Child* 2011; 96(11):998–1002
- 12 Benini F, Ferrante L, Pozza VD, Trapanotto M, Facchin P. Children's needs: key figures from the Veneto region, Italy. *Eur J Palliat Care* 2008;15(06):299–304
- 13 Gans D, Kominski GF, Roby DH, et al. Better outcomes, lower costs: palliative care program reduces stress, costs of care for children with life-threatening conditions. *Policy Brief UCLA Cent Health Policy Res* 2012;2012–3(PB2012-3):1–8
- 14 Keele L, Keenan HT, Sheetz J, Bratton SL. Differences in characteristics of dying children who receive and do not receive palliative care. *Pediatrics* 2013;132(01):72–78
- 15 Postier A, Chrastek J, Nugent S, Osenga K, Friedrichsdorf SJ. Exposure to home-based pediatric palliative and hospice care and its impact on hospital and emergency care charges at a single institution. *J Palliat Med* 2014;17(02):183–188
- 16 Kassam A, Skiadaresis J, Alexander S, Wolfe J. Parent and clinician preferences for location of end-of-life care: home, hospital or freestanding hospice? *Pediatr Blood Cancer* 2014;61(05):859–864