

Positive impact on children's quality of life under non-invasive home mechanical ventilation – PedsQL application in a monocentric cohort

Ana Moura-Figueiredo^{1a*}, Joana Amara^{2b}, Guiomar Oliveira^{3,4c}, Núria Madureira^{5d}, Miguel Félix^{5e}, and Cândida Cancelinha^{2,3,6f}

¹Pediatrics Department, Centro Hospitalar do Médio Tejo, Torres Novas; ²Hospital Pediátrico, Centro Hospitalar e Universitário de Coimbra; ³Faculty of Medicine, University of Coimbra; ⁴Research and Clinical Training Center, Hospital Pediátrico do Centro Hospitalar e Universitário de Coimbra; ⁵Pediatric Pulmonology Unit, Hospital Pediátrico do Centro Hospitalar e Universitário de Coimbra; ⁶Pediatric Palliative Care Team, Hospital Pediátrico do Centro Hospitalar e Universitário de Coimbra. Coimbra, Portugal

ORCID: ^a0000-0003-3463-0193; ^b0000-0003-1493-7892; ^c0000-0002-7049-1277; ^d0000-0002-4738-9880; ^e0000-0003-2973-0413;

^f0000-0001-8816-5990

Abstract

Introduction: In home mechanical ventilated (HMV) children, treatment is not based on cure, but rather on preventing morbidity and mortality and promoting their well-being. This study aimed to evaluate the different dimensions of quality of life (QoL), of HMV children/adolescents of a tertiary pediatric hospital, according to children/adolescents and caregiver's perspective, and relating it to sociodemographic and clinical characteristics. **Methods:** We studied families who integrated the home-ventilation program. The PedsQL™ questionnaire was answered by children/adolescents and their caregivers. Sociodemographic and clinical information was recorded. **Results:** 46 parents and 39 children/adolescents' questionnaires were answered. The median of the average duration of ventilatory support was 3,5y. 60,9% attend full-time school. The average of general QoL, according to parents and children/adolescents, was 61,6 and 64,0, respectively, with a strong correlation between two groups ($r = 0,691$). Children/adolescents showed higher scores for all dimensions of QoL. In the physical dimension a very strong correlation between groups was verified. For both groups it was found that psychosocial dimension is the best predictor of QoL. **Discussion/conclusion:** Parents and children/adolescents shared the same perspective about QoL, with a good correlation between answers of both groups, concluding that home ventilation is an acceptable medical strategy and has impact in QoL for both.

Keywords: Quality of life. Non-invasive ventilation. Children. Adolescents.

Impacto positivo na qualidade de vida de crianças sob ventilação mecânica não invasiva domiciliar – aplicação PedsQL em uma coorte monocêntrica

Resumo

Introdução: Nas crianças dependentes de ventilação crónica no domicílio, o tratamento não tem por base a cura, mas sim a prevenção da morbimortalidade e promoção do seu bem-estar. Este estudo pretendeu avaliar as diferentes dimensões da qualidade de vida (QV), em crianças/adolescentes sob ventilação mecânica domiciliária de um hospital

*Correspondence:

Ana Moura-Figueiredo

E-mail: anammouraf@gmail.com

Received: 18-03-2022

Accepted: 15-02-2023

<https://pjp.spp.pt>

Available online: 04-08-2023

Port J Pediatr. 2023;54(3):147-155

DOI: 10.24875/PJP.M23000004

2184-3333 / © 2023 Portuguese Society of Pediatrics. Published by Permalyer. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

pediátrico terciário, de acordo com a perspectiva dos próprios e dos seus cuidadores, relacionando-a com características sociodemográficas e clínicas. **Métodos:** Foram estudadas famílias que integravam um programa de ventilação no domicílio. O questionário PedsQL™ foi respondido por crianças/adolescentes e seus cuidadores. Foram recolhidas variáveis clínicas e sociodemográficas. **Resultados:** Responderam 46 pais e 39 crianças/adolescentes. A mediana do tempo de suporte ventilatório foi 3,5 anos. 60,9% frequentaram a escola a tempo inteiro. As médias da QV geral segundo pais e crianças/adolescentes foram 61,6 e 64,0, respetivamente, com forte correlação entre os dois grupos ($r = 0,691$). As crianças/adolescentes apresentaram scores mais elevados para todas as dimensões de QV. Na dimensão física verificou-se uma forte correlação entre os grupos. Para ambos os grupos constatou-se que a dimensão psicossocial é o melhor preditor de QV. **Discussão/conclusão:** Pais e crianças/adolescentes partilham a mesma perspetiva sobre a QV, com boa correlação entre as respostas, concluindo-se que a ventilação no domicílio é uma estratégia médica aceitável e com impacto na QV para ambos.

Palavras chave: Qualidade de vida. Ventilação não invasiva. Crianças. Adolescentes.

Keypoints

What is known

- In home mechanical ventilated (HMV) children, treatment is not based on cure, but rather on preventing morbidity and mortality and promoting their well-being.
- Since HMV became an option, in the beginning of 80's, several studies appeared, evaluating its medical and economical components, but few focused in patients QoL.

What is added

- To demonstrate that in general, parents and children/adolescents share the same positive, with good correlation between answers.
- Home ventilation is an acceptable medical strategy and has impact in QoL for both, parents and the children/adolescents.

Introduction

In the last decades there has been a progressive increase in the number of children in need for prolonged mechanical ventilation¹⁻³. This growth, along with the increasing pressure for hospital discharge and cost reduction, has led to the establishment of home mechanical ventilation programs^{1,4}. For children in need for chronic home mechanical ventilation (HMV) the treatment goal is not a cure, but instead mortality and morbidity prevention as well as well-being promotion. By reducing morbidity, the child and its family's quality of life (QoL) increases, allowing a better social reintegration⁴.

In 1993, for the first time in Portugal, a HMV program for chronic respiratory insufficiency patients was established in Coimbra's University Pediatric Hospital^{1,4,5}. Since then, two hundred children and adolescents took part in this program. Health related QoL (HRQoL) is a subjective, multidimensional concept encompassing many elements such as physical, psychologic, social and behavioral wellbeing and function as perceived by individuals^{6,7}. QoL evaluation has been increasingly important as a fundamental measure to corroborate health interventions' efficacy⁷. Since HMV became an option, in the beginning of the 80's, several studies appeared, evaluating its medical and economical components, but few focused in patients QoL⁸. Most studies describing impact of ventilation refer to adults

or to larger groups that included children, but those who focused only on the latter were scarce⁹. In Portugal there is not any published study approaching the impact of HMV in children and adolescents' QoL.

In this study, authors intend to evaluate QoL in children and adolescents under non-invasive HMV, according to their own and their caregivers' perspective in a university pediatric hospital (group III) in the Center Region of Portugal, to understand which dimensions (physical, emotional, social and scholar) are most influenced in QoL and to relate it with sociodemographic and clinical features of patients.

Methods

Sample characterization

A cross-sectional cohort study was performed with families integrated in a level III university pediatric hospital home ventilation program, for a period of 8 months (from June 2016 to January 2017).

Inclusion criteria were the following: children/adolescents under home non-invasive ventilation (NIV) for at least six months, aged two or more years of age (at the time of study period). In children between two and eight years of age, the questionnaire was answered by their parents. Informed consent was obtained from their parents in all situations.

Evaluation tool characterization

QoL questionnaire application, Portuguese version of "Pediatric Quality of Life Inventory™ (PedsQL™)"¹⁰, self-filling, anonymous, with answers based on the Likert scale, with five minutes estimated filling time. Consent for this questionnaire use was obtained through the online platform ePROVIDE™.

PedsQL™ is a generic tool for measuring, monitoring and evaluating HRQoL in healthy children and adolescents, as well as with those in acute or chronic conditions. It is comprised of 23 items, separated in four dimensions: physical functionality (8 items), emotional functionality (5 items), social functionality (5 items) and scholar functionality (5 items). Questions are related to a problem in the last month, scored in a scale of 0 to 4, where 0 represents "it is never a problem" and 4 "it is always a problem". This score was posteriorly converted in a scale of 0 to 100 (0 = 100, 1 = 75, 2 = 50, 3 = 25 and 4 = 0). Higher scores indicate higher QoL. In the end, adding each dimension items (in the 0-100 scale) and dividing by the number of items that each other has, a specific QoL value was obtain for each dimension. The overall QoL value is achieved adding the 4 dimensions totals and dividing by 4. Beyond these dimensions, a psychosocial dimension was also created by adding the emotional, social and scholar totals and dividing by 3. Dimensions lacking more than 50% answered of the items were excluded.

This tool is applicable to children and adolescents between the age of 2 and 18 years, in two formats: one self-filling for the children and one to be filled by the parents about their child. The self-filling questionnaire for children was applied in the ages between 8 and 12 years and 13 and 18 years, all of which with answer ability. The parent-filling questionnaire was applied in the ages between 2 and 4 years (very small children), 5 and 7 years (small children), 8 and 12 years (children) and 13 and 18 years (adolescents).

The questionnaire and informed consent were delivered to the children/adolescents and their caregivers by the nursing team that carried the pre-appointment triage, filled anonymously, and returned, in a sealed envelope, to the consulting physician that afterwards delivered them to the research team.

Sociodemographic variables were collected, as age, gender and school attendance (full time, partial time or no attendance), clinical variables, ventilation support time, ventilation mode (continuous positive airway pressure – CPAP – or bilevel positive airway pressure – BiPAP), type of interface (nasal, oro-nasal or facial

mask), average daily ventilation duration (between 0 to 8 hours, 8 to 16 hours or more than 16 hours), ventilation compliance considering the number of nights under ventilation registered in the ventilator electronic record (classified as insufficient if less than 50%, acceptable if 50 to 80% or good/very good if more than 80%), primary diagnosis and associated comorbidities. Primary diagnoses that prompted the beginning of the ventilation were organized into seven groups: central hypoventilation, airway obstruction, chronic pulmonary disease, neuromuscular disease, genetic syndrome, hereditary metabolic disease and neurologic disease.

Statistical analysis

Obtained data was processed with *Microsoft Office Excel 2013*® and their inferential and descriptive analysis were carried with *Statistical Package for the Social Sciences*® software (SPSS®, version 24.0). Statistical tests were evaluated using a 5% significance level.

Quantitative variables normality was tested using *Shapiro-Wilk* and *Kolmogorov-Smirnov* tests. Qualitative variables values were displayed according to their absolute and relative frequency, as quantitative variables values were displayed by median, inter-quartile range and extremes (maximum and minimum) for the age and ventilatory support duration variables and by average $\pm$ standard deviation or median for QoL related variables.

General QoL according to parents was compared with general QoL according to children and was posteriorly grouped into three levels of life quality – low (0-33), medium (34-66) and high (67-100) – according to the results obtained through PedsQL™. Values according to parents obtained in each dimension (physical, emotional, social, scholar and psychosocial) were also compared with values according to children, for each respective dimension.

To analyze a potential correlation between the values according to parents and children, Pearson coefficient was used for normal distribution variables and Spearman coefficient for non-normal distribution variables.

Comparison between general QoL, according to parents and children, and average daily ventilation time (0-8 hours and 8-16 hours) was made using t-Student test for independent samples.

To compare general QoL, according to parents and children, with ventilation type (CPAP and BiPAP) and interface type (nasal and oro-nasal), Mann-Whitney U test was used.

General QoL, according to parents and children, was also compared with ventilation compliance using the

Kruskal-Wallis test. *Post-hoc* testing was performed to compare averages between each pair of groups, using Dunn correlation for multiple comparisons.

Spearman coefficient was used to analyze a potential correlation between QoL, according to parents and children and average ventilation time.

To compare different dimensions related to QoL (physical, emotional, social, scholar and psychosocial), according to parents and children, with average daily ventilation time, parametric tests were used for normal distribution variables, t-Student test for independent samples and non-parametric tests for non-normal distribution variables, Mann-Whitney U test.

To establish a comparison between the different studied dimensions with ventilation type, interface type and ventilation compliance, non-parametric tests were used, namely Mann-Whitney U test for ventilation type and interface type and Kruskal-Wallis test for ventilation compliance. When using the latter, *post-hoc* testing was carried to compare averages between each pair of groups, using Dunn correlation for multiple comparisons.

To test the influence that physical and psychosocial dimension have in general QoL, according to parents and children, linear regression analysis was completed using the Enter method.

Results

During the study period, 46 children and adolescents undergoing non-invasive home ventilation were included, representing 60,5% of children/adolescents followed in the Pulmonology clinic at a pediatric tertiary hospital, that met the inclusion criteria. A total of 46 parents and 39 children answered the questionnaire, with a 100% response rate. Seven children did not answer due to their age (under 8 years-old). Clinical and sociodemographic variables are shown in Table 1.

Results obtained for general QoL between Parents and children/adolescents scores

Fig. 1 shows the results of the comparison between general QoL according to parents and children/adolescents, mean score $\pm$ SD respectively ($64,0 \pm 21,3$; $61,6 \pm 17,6$). There was a statistically significant and strong correlation between the two scores obtained ($r = 0,691$, $p < 0,001$).

Diagnostic group

No inferential analysis was performed due to the small sample size. Therefore, no differences were

Table 1. Sociodemographic and clinical variables of children enrolled in the study

Number of patients	46, male 26 (56,5%)
Patient age range	Median 13 years-old, IQR 9-16 years-old, range 2-19 years-old 2-4 years-old: 5 (10.9%) 5-7 years-old: 2 (4.3%) 8-12 years-old: 14 (30.4%) ≥ 13 years-old: 25 (54.3%)
School attendance	Full time: 28 (60.9%) Partial time: 9 (19.6%) No attendance: 5 (10.9%)
Ventilatory support time	Median 3,5 years, IQR 1.8-6.2 years, range 0.5-17 years
Ventilation mode	BiPAP: 37 (80.4%) CPAP: 9 (19.6%)
Interface type	Oro-nasal: 35 (76.1%) Nasal: 11 (23.9%) Facial: 0 (0%)
Daily ventilation duration	0-8 h: 29 (63%) 8-16 h: 17 (37%) > 16 h: 0 (0%)
Compliance	Good/very good: 32 (69.6%) Acceptable: 10 (21.7%) Insufficient: 4 (8.7%)
Diagnostic group	Neuromuscular disease: 14 (30.4%) Airway obstruction: 13 (28.3%) Genetic syndrome: 10 (21.1%) Chronic pulmonary disease: 4 (8.7%) Central hypoventilation: 3 (6.5%) Metabolic disease: 1 (2.2%) Neurologic disease: 1 (2.2%)

BiPAP: bilevel positive airway pressure; CPAP: continuous positive airway pressure; IQR: interquartile range.

found between diagnostic groups. Scores results are showed in Table 2.

Average daily ventilation time

General QoL of children/adolescents with ventilation between 0-8 hours daily duration was found not to be significantly different from children/adolescents with 8-16 hours daily duration, neither for parents nor children/adolescents ($p = 0.295$ and $p = 0.693$) (Table 3).

Mode of ventilation

General QoL, according to parents and children/adolescents, was not significantly different depending on

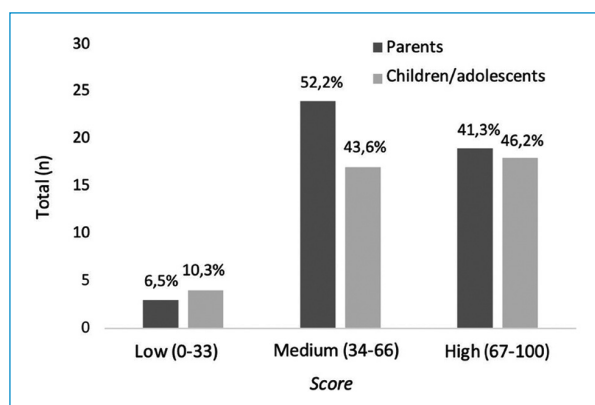


Figure 1. General QoL according to parents and children/adolescents.

Table 2. Quality of life to parents and children/adolescents, according to diagnostic groups

Diagnostic groups	General QoL (Parents)	General QoL (children/adolescents)
Neuromuscular disease (n = 14)	55.0	62.3
Airway obstruction (n = 13)	63.7	70.8
Genetic syndrome (n = 10)	59.8	57.5
Chronic pulmonary disease (n = 4)	76.4	80.2
Central hypoventilation (n = 3)	81.7	83.1
Metabolic disease (n = 1)	51.1	23.4
Neurologic disease (n = 1)	53.0	53.0

the mode of ventilation used (CPAP or BiPAP) ($p = 0.070$ and $p = 0.242$) (Table 3).

Type of interface

It was found that the general QoL, according to parents, with oro-nasal interface was not significantly different from the general QoL with nasal interface ($p = 0.867$) (Table 3). According to children and adolescents, it was found that general QoL was significantly lower with oro-nasal interface than with nasal interface (median 65.8 vs 87.0, respectively, $p = 0.036$) (Table 3).

Ventilation compliance

Regarding general QoL, comparing between parents and children/adolescents, there was a statistically significant difference with ventilation compliance ($p = 0.019$ and $p = 0.044$) (Table 3). For both, there is a higher QoL with good/very good compliance when compared with insufficient compliance (median 62.6 vs 31.3 for parents and median 70.6 vs 40.3 for children/adolescents) (Table 3).

Correlation with ventilatory support duration

Assessing the general QoL according to the duration of ventilatory support, a positive and weak correlation was established ($p = 0.341$) between the variables for parents (Fig. 2). Despite the positive correlation between QoL and ventilatory support duration in children/adolescents, it had a very weak correlation ($p = 0.191$) without statistical significance ($p = 0.245$) (Fig. 2).

Results obtained for dimensions of general QoL between

Parents and children/adolescents scores

A comparative analysis was done between all dimensions of general QoL, according to parents and children/adolescents, with globally higher scores in all dimensions for children/adolescents' group. The comparison between different dimensions of QoL, according to the perspective of parents and children/adolescents, showed that a general QoL increase follows an increase in all dimensions ($r = 0.833$ for physical dimension, $r = 0.547$ for emotional dimension, $p = 0.492$ for social dimension and $p = 0.546$ for scholar dimension).

Diagnostic group

No inferential analysis was performed due to the small sample size.

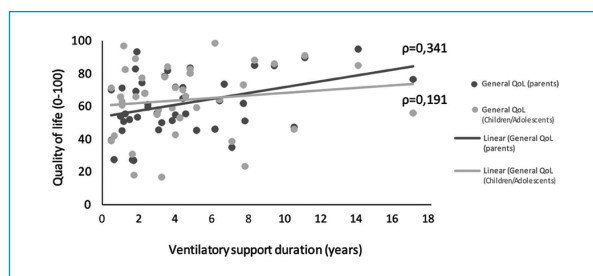
Average daily ventilation duration

According to parents and children/adolescents, there were no significant differences between 0-8 hours versus 8-16 hours for the physical, emotional, social and scholar dimensions ($p \geq 0.05$) (Table 4).

Table 3. Quality of life, according to parents and children/adolescents, according to average daily ventilation duration, mode of ventilation, type of interface and ventilation compliance type

	General QoL (parents) and range	p-value (parents)	General QoL (C/A) and range	p-value (C/A)
Average daily ventilation duration 0-8h (n = 29) 8-16h (n = 17)	55,5 (27,0-95,0) 69,8 (39,4-89,8)	p = 0,295	65,9 (16,7-98,9) 62,0 (23,4-91,1)	p = 0,693
Ventilation mode BiPAP (n = 37) CPAP (n = 9)	55,5 (27,0-95,0) 71,7 (50,8-84,8)	p = 0,070	64,9 (16,7-98,8) 71,4 (42,7-96,9)	p = 0,242
Type of interface Oro-nasal (n = 35) Nasal (n = 11)	61,9 (27,0-89,8) 48,4 (45,4-95,0)	p = 0,867	65,8 (16,7-91,1) 87,0 (58,0-98,8)	p = 0,036
Ventilation compliance type Good/very good (n = 32) Acceptable (n = 10) Insufficient (n = 4)	62,6 (27,3-95,0) 69,2 (45,2-82,8) 31,3 (27,0-51,3)	p = 0,019	70,6 (23,4-98,8) 62,8 (16,7-89,1) 40,3 (18,0-59,1)	p = 0,044

C/A: children/adolescents.

**Figure 2.** Correlation between general quality of life and ventilatory support duration.

Mode of ventilation

According to parents, there were statistically significant differences between CPAP and BiPAP, for the physical and social dimensions (median 74.6, $p = 0.016$ and median 80.6, $p = 0.015$, respectively) (Table 4). To children/adolescents there were no statistically significant differences for any dimensions ($p \geq 0.05$) (Table 4).

Type of interface

For every studied QoL dimensions, according to parents, there were no statistically significant differences between interface types ($p \geq 0.05$) (Table 4). According to the children/adolescents, and considering the scholar dimension, there was a statistically significant difference between interface types, with higher values in nasal interface (median 84.0 vs 57.2, $p = 0.012$) (Table 4).

Ventilation compliance type

For the parents there were statistically significant differences between physical and emotional dimensions between children/adolescents with insufficient and good/very good compliance (median 25.8 vs 61.1, $p = 0.042$ and median 32.5 vs 65.3, $p = 0.048$, respectively) (Table 4). For other dimensions (social and scholar) there were not statistically significant differences ($p \geq 0.05$) (Table 4). According to children/adolescents' answers, there were statistically significant differences between insufficient and good/very good compliance to physical dimension ($p = 0.029$) (Table 4).

Psychosocial dimension

As for psychosocial dimension, comparing with other studied dimensions, according to parents, only the difference between ventilation compliance type was statistically significant [$\chi^2 (2) = 6.433$, $p = 0.040$], namely between insufficient and good/very good compliance (35.3 vs 64.3, $p = 0.037$). There were no statistically significant differences between insufficient and acceptable compliance ($p = 0.077$) and acceptable and good/very good compliance ($p = 1.000$). According to children and adolescents, in psychosocial dimension, there was a higher QoL with nasal interface (median 65.0) than oro-nasal interface (median 88.3) ($U = 43.5$; $p = 0.031$).

Table 4. Quality of life dimensions, according to parents and children/adolescents, according to average daily ventilation duration, mode of ventilation, type of interface and ventilation compliance type

	Physical dimension		Emotional dimension		Social dimension		Scholar dimension	
	General QoL (mean)							
	P	C/A	P	C/A	P	C/A	P	C/A
Average daily ventilation duration								
0-8h (n = 29)	53,3	58,5	58,8	66,0	57,2	67,4	52,1	62,8
8-16h (n = 17)	65,8	63,0	65,3	62,9	75,3	63,8	60,3	56,7
p-value	0,905	0,874	0,501	0,558	0,061	0,633	0,894	0,543
Ventilation mode								
BiPAP (n = 37)	54,2	55,8	61,2	64,8	60,3	62,7	55,1	59,8
CPAP (n = 9)	74,6	77,7	61,9	65,7	80,6	81,4	56,3	65,0
p-value	0,016	0,090	0,901	0,898	0,015	0,102	0,850	0,545
Type of interface								
Oro-nasal (n = 35)	58,5	58,0	62,3	62,8	64,8	63,1	55,8	57,2
Nasal (n = 11)	56,9	72,5	57,8	79,0	62,2	86,0	53,5	84,0
p-value	0,911	0,331	0,660	0,089	0,632	0,058	0,761	0,012
Ventilation compliance type								
Good/very good (n = 32)	61,1	65,6	65,3	68,5	66,6	69,4	57,0	64,4
Acceptable (n = 10)	63,7	60,3	61,3	60,7	64,4	62,9	59,4	62,9
Insufficient (n = 4)	25,8	22,7	32,5	50,0	47,5	51,3	35,0	33,8
p-value	0,042	0,029	0,048	0,145	0,292	0,418	0,116	0,121

P: parents; C/A: children/adolescents.

Comparison between physical and psychosocial dimensions with general QoL, according to parents and children/adolescents scores

The results showed that both physical and psychosocial dimensions influence the general QoL, despite psychosocial dimension showing higher correlation score for both parents and children/adolescents ($\beta = 0.703/\beta = 0.793$ vs $\beta = 0.391/\beta = 0.333$).

Discussion

HRQoL has been increasingly acknowledged as a significant result in pediatric health care practice¹¹. Therefore, in the last decade there has been a sharp increase in development and use of HRQoL evaluation tools in pediatric age in order to improve children's health and well-being and to determine provided services' efficacy^{11,12}. To this date, many studies focused on QoL analysis in adults with several chronic diseases but there is a relative absence of similar studies dedicated to pediatrics^{9,12}.

Home mechanical ventilation use was hypothetically associated with lower HRQoL levels in pediatric age¹³ and health care professionals initially underestimated QoL in children with serious conditions^{14,15}.

Some existing studies involving children in need of chronic home ventilation evaluated QoL through questionnaires filled only by the parents¹³, since children can be considered unreliable, and for that reason, initial attempts to evaluate children HRQoL were based in data given mainly by mothers¹⁶. Parents mostly rate their child's QoL lower than children/adolescents themselves¹⁷.

In our study, comparing both perspectives, general QoL median score, according to parents and children/adolescents, is very similar, as described by a Canadian study¹⁸. When comparing general QoL results with previous studies using PedsQLTM, we can conclude that the scores of our study were superior to those obtained by a Canadian study (parents 35,4/children 33,2)¹⁸ and similar to a Spanish study (parents 52,1/children 61,4)¹⁹, as approximately 90% of children/adolescents participating in our study reported a general QoL score medium or high. In a Swedish study, where a similar questionnaire was used, scores of HRQoL obtained by parents were comparable to our study²⁰.

Other studies showed higher general QoL values for healthy children/adolescents (averaging 76.8-88.9) when comparing QoL scores obtain in the current study (averaging 64.0 by children/adolescents and 61.2 by parents)^{9,11}. These findings were also confirmed in a recently published revision article¹⁷.

In our study, the majority (80.5%) of school age children/adolescents attend school regularly, similarly to a Canadian study (more than 80%)¹⁸. Besides, in our sample, 60.9% attend school in a full-time regime, higher than the mentioned study (46.7%)¹⁸.

Concerning different diagnostic groups, an American study concluded that there was not any statistically significant differences between disease types⁸. In our study, no inferential analysis was performed due to the small sample size. Two Malaysian and Spanish studies^{19,21} showed a superior general QoL score in respiratory tract disease groups. Interestingly, QoL score was almost always superior when answered by children/adolescents comparing to parents, similarly to a Spanish study¹⁹ and contrasting with a Canadian study¹⁸, where the authors used the same questionnaire.

No publications were found that compared general QoL between average daily ventilation, type of ventilation, type of interface and compliance to ventilation.

According to parents, general QoL was correlated with ventilatory support duration (in years), unlike what was found in the Canadian study¹⁸.

The biggest difference in perspective was found in the schooling dimension, similarly to an English study⁹. Physical dimension showed the strongest correlation between obtained scores, as stated by a Brazilian study¹¹, and, on the contrary, social dimension showed the weakest, which shows that parents and children/adolescents concur more in the influence of physical component in children/adolescents' QoL. Other studies describe physical dimension as the one with the lowest scores, which is corroborated by our study in children's response^{19,22}, approaching the score obtained in scholar dimension.

Ventilation mode and ventilation compliance, according to parents, was what influenced children/adolescents' QoL at a physical level. To them, social and emotional QoL are also influenced by these variables, respectively.

When comparing average scores in isolated dimensions with psychosocial component's average score, it is possible to conclude that emotional and social components have a more positive influence in psychosocial dimension than scholar dimension, which shows lower average scores.

When evaluating which dimension, physical or psychosocial, has more impact in general QoL, parents and children/adolescents agree that psychosocial has more relevance. This is also observed in a study enrolling children with Duchenne's Muscular Dystrophy, showing that disease may affect family functioning more severely in psychosocial terms, than in physical terms²². These

data suggest the importance of multi-disciplinary approach to children with respiratory chronic disease, namely psychological support.

One limitation of this study is the small sample size obtained which, although we find them representative of the followed patient group in consultation, limits some results interpretation.

The fact that questionnaires were filled out before the consultation and delivered, even in a sealed envelope, to the responsible physician, may constitute a bias in the type of answers obtained.

The instrument used (PedsQL™) is not disease specific and therefore does not provide detailed information on specific factors that vary according to the underlying pathology and may influence QoL in chronically ventilated children/adolescents at home. To our knowledge, there is no known validated Portuguese QoL questionnaire related to specific complex chronic diseases.

In conclusion, this study showed that, in general, parents and children/adolescents share the same perspective, with a good correlation between answers, concluding that home ventilation is an acceptable medical strategy and has impact in QoL for both.

Even though children/adolescents under chronic home ventilation show lower QoL scores when compared with healthy children's, surprisingly, they are not as low as expected.

In future studies, determining children and adolescents' QoL before starting home ventilation would be important to understand the impact of these programs in QoL. Considering that chronic illness is a potential source of family stress, further assessing caregivers' QoL is also recommended.

Acknowledgments

To all the pediatric staff that contributed to the project and a special recognition to every parent and child involved in the study, for their availability and prompted answering to the questionnaires.

Funding

None.

Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with

the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

References

1. Cancelinha C, Madureira N, Mação P, et al. Long-term ventilation in children: Ten years later. *Rev Port Pneumol*. 2015;21(1):16-21. doi:10.1016/j.rppnen.2014.03.017
2. Ottonello G, Ferrari I, Pirroddi IMG, et al. Home mechanical ventilation in children: Retrospective survey of a pediatric population. *Pediatr Int*. 2007;49(6):801-805. doi:10.1111/j.1442-200X.2007.02463.x
3. Resener TD, Martinez FE, Reiter K, Nicolai T. Assistência ventilatória domiciliar em crianças - descrição de um programa. *J Pediatr (Rio J)*. 2001;77(2):84-88. http://www.jpmed.com.br/conteudo/01-77-02-84/port_print.htm
4. Estêvão M. Ventilação não invasiva no domicílio em pediatria. *Acta Pediatr Port*. 2000;31(2):135-141.
5. Maia C, Gomes AC, Silva TR, Félix M, Estêvão MH. Apoio ventilatório em doentes neuromusculares - experiência do Hospital Pediátrico de Coimbra. *Acta Pediatr Port*. 2011;42(1):12-16. doi:0873-9781/11/42-1/12
6. Ravens-Sieberer U, Gosch A, Abel T, et al. Quality of life in children and adolescents: a European public health perspective. *Soz Präventivmed*. 2001;46(5):294-302.
7. Iannaccone ST, Hynan LS, Morton A, et al. Neuromuscular Disorders The PedsQL TM in pediatric patients with Spinal Muscular Atrophy: Feasibility, reliability, and validity of the Pediatric Quality of Life Inventory TM Generic Core Scales and Neuromuscular Module q. *Neuromuscul Disord*. 2009;19(12):805-812. doi:10.1016/j.nmd.2009.09.009
8. Lumeng JC, Warschausky SA, Nelson VS, et al. The quality of life of ventilator-assisted children. *Pediatr Rehabil*. 2001;4(1):21-27. doi:10.1080/13638490151068438
9. Noyes J. Comparison of ventilator-dependent child reports of health related quality of life with parent reports and normative populations. *J Adv Nurs*. 2007;1(58):1-10. doi:10.1111/j.1365-2648.2006.04191.x
10. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. *Med Care*. 2001;39(8):800-812. doi:10.1097/00005650-200108000-00006
11. Klatchoian D a, Len C a, Terrerri MTR a, et al. Quality of life of children and adolescents from São Paulo: reliability and validity of the Brazilian version of the Pediatric Quality of Life Inventory version 4.0 Generic Core Scales. *J Pediatr (Rio J)*. 2008;84(4):308-315. doi:10.2223/JPED.1788
12. Varni JW, Limbers CA, Burwinkle TM. Impaired health-related quality of life in children and adolescents with chronic conditions: a comparative analysis of 10 disease clusters and 33 disease categories/severities utilizing the PedsQLTM 4.0 Generic Core Scales. *Health Qual Life Out-comes*. 2007;5(43). doi:10.1186/1477-7525-5-43
13. Mah JK, Thannhauser JE, Kolski H, Dewey D. Parental Stress and Quality of Life in Children With Neuromuscular Disease. *Pediatr Neurol*. 2008;39(2):102-107. doi:10.1016/j.pediatrneurol.2008.04.011
14. Kohler M, Clarenbach CF, Böni L, Brack T, Russi EW, Bloch KE. Quality of life, physical disability, and respiratory impairment in Duchenne muscular dystrophy. *Am J Respir Crit Care Med*. 2005;172(8):1032-1036. doi:10.1164/rccm.200503-322OC
15. Bach JR, Vega J, Majors J, Friedman A. Spinal muscular atrophy type 1 quality of life. *Am J Phys Med Rehabil*. 2003;82(2):137-142. doi:10.1097/01.PHM.0000046622.57204.05
16. Eiser C, Morse R. A review of measures of quality of life for children with chronic illness. *Arch Dis Child*. 2001;84:205-211. doi:10.1136/a-dc.84.3.205
17. Mattson J, Lunnelie J, Löfholm T, Andersson ES, Aune RE, Björling G. Quality Of Life in Children With Home Mechanical Ventilation – A Scoping Review. *SAGE Open Nurs*. 2022;8:1-12. doi:10.1177/23779608221094522
18. Seear M, Kapur A, Wensley D, Morrison K, Behrooz A. The quality of life of home-ventilated children and their primary caregivers plus the associated social and economic burdens: a prospective study. *Arch Dis Child*. 2016;0:1-8. doi:10.1136/archdischild-2015-309796
19. González R, Bustinza A, Fernandez SN, et al. Quality of life in home ventilated children and their families. *Eur J Pediatr*. 2017;176(10):1307-1317. doi:10.1007/s00431-017-2983-z
20. Israelsson-Skogsberg Å, Persson C, Markström A, Hedén L. Children with home mechanical ventilation—Parents' health-related quality of life, family functioning and sleep. *Acta Paediatr Int J Paediatr*. 2020;109(9):1807-1814. doi:10.1111/apa.15177
21. Tan LT, Nathan AM, Jayanath S, et al. Health-related quality of life and developmental outcome of children on home mechanical ventilation in a developing country: A cross-sectional study. *Pediatr Pulmonol*. 2020;55(12):3477-3486. doi:10.1002/ppul.25083
22. Baiardini I, Minetti C, Bonifacino S, et al. Quality of life in Duchenne muscular dystrophy: the subjective impact on children and parents. *J Child Neurol*. 2011;26(6):707-713. doi:10.1177/0883073810389043