Effects of noninvasive ventilation on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis: Randomized controlled trial

Cibelle Andrade Lima a, Armèle de Fátima Dornelas de Andrade b,*, Shirley Lima Campos b, Daniella Cunha Brandão b, Guilherme Fregonezi c, Ianny Pereira Mourato b, Andrea Aliverti d, Murilo Carlos Amorim de Britto e

a Universidade Federal de Pernambuco — UFPE, Rua Pessoa de Melo, 333, 702, Madalena, Recife, PE 50721610, Brazil
b Universidade Federal de Pernambuco — UFPE, Departamento de Fisioterapia, Av. Jornalista Aníbal Fernandes, Cidade Universitária, Recife, PE 50740-560-901, Brazil
c Departamento de Fisioterapia, Universidade Federal do Rio Grande do Norte, Campus Universitário Lagoa Nova, Caixa Postal 1524, CEP:59072-970 Natal-RN, Brazil
d Dipartimento di Elettronica, Informazione e Bioingegneria Politecnico di Milano, Piazza Leonardo da Vinci, 32, I-20133 Milan, Italy
e Instituto Materno Infantil de Pernambuco, Rua dos Coelhos 300, Pós Graduação, Boa Vista, Mailbox: 1393, Recife, PE 50070-550, Brazil

Received 5 December 2013; accepted 9 April 2014
Available online 23 April 2014

KEYWORDS
Cystic fibrosis; Exercise tolerance; Exercise test; Noninvasive ventilation

Summary
Background: Dyspnea and exercise intolerance are the symptoms that most affect the quality of life of children and adolescents with respiratory disorders resulting from cystic fibrosis (CF).
Objective: To evaluate the effect of noninvasive ventilation (NIV) on treadmill 6-min walk distance and regional chest wall volumes in cystic fibrosis patients.
Method: Crossover clinical trial, randomized, controlled and open with 13 children and adolescents with CF, aged 7–16 years, with pulmonary impairment (NTC01987271). The patients...
performed a treadmill walking test (TWT) during 6 min, with and without NIV on a BiLEVEL mode, an interval of 24–48 h between tests. Before and after each test, patients were assessed by spirometry and optoelectronic plethysmography.

**Results:** Walking distance in TWT with NIV was significantly higher that without ventilatory support (mean ± sd: 0.41 ± 0.08 vs. 0.39 ± 0.85 km, p = 0.039). TWT with NIV increase forced expiratory volume on 1 s (FEV₁; p = 0.036), tidal volume (Vt; p = 0.005), minute ventilation (MV; p = 0.013), pulmonary rib cage volume (Vrcp; p = 0.011), and decrease the abdominal volume (Vab; p = 0.013) after test. There was a significant reduction in oxygen saturation (p = 0.018) and permanent increase in respiratory rate after 5 min (p = 0.021) after the end test without NIV.

**Conclusion:** During the walking test on the treadmill, the NIV change thoracoabdominal kinematics and lung function in order to optimized ventilation and tissue oxygenation, with improvement of walk distance. Consequently, NIV is an effective tool to increase functional capacity in children and adolescents with cystic fibrosis.

© 2014 Elsevier Ltd. All rights reserved.

---

**Introduction**

Cystic fibrosis (CF) is a multisystemic disorder in which the respiratory system is affected in 95% of the cases, and the primary cause of death in young adults with the disease [1].

Pulmonary impairment is progressive with recurring infectious and inflammatory processes of varying intensity, involving symptoms such as hypersecertivity and production of thick tracheobronchial mucus, peripheral airway obstruction, signs of hyperinflation associated or not to collapsed lung and atelectasis. These processes culminate in dyspnea and exercise intolerance, symptomatology that limits activities of daily living and interferes with the quality of life of cystic fibrosis patients [1–4].

In addition to respiratory limitations, other factors also account for reduced functional capacity (FC), including peripheral muscle weakness, systemic inflammation, oxidative stress, malnutrition and the inactivity. These factors are a vicious cycle for deteriorated aerobic conditioning and increased mortality levels in this population [5–8].

Even though there is still no curative treatment, survival rates have been increasing due to early diagnosis, multiprofessional management in specialized centers and access to adequate therapy, mainly in the control of pulmonary infections, adequate nutrition and as normal a lifestyle as possible. In this respect, there is a need to investigate strategies that facilitate physical activities, improving physical conditioning and exercise tolerance in this population [9,10].

Noninvasive ventilation (NIV) has been used to treat patients with acute and chronic respiratory failure. Studies with chronic pulmonary patients, including those with cystic fibrosis, show the benefits of using on alveolar ventilation, gas exchange, reduced air trapping and pulmonary hyperinflation. It is also an effective instrument in improving ventilatory pattern and reducing respiratory work. However, the immediate effect of this intervention on daily activities and functional capacity in cystic fibrosis patients remains unknown and encourages this research [6,11–20].

Thus, the aim of the present study was to assess the effect of using NIV during the treadmill walking test on walk distance, as well as the cardiovascular and thoracoabdominal kinematic variables of children and adolescents with CF.

The primary outcome variable is walk distance (WD) in the TWT and the secondary outcomes are peripheral O₂ saturation (SpO₂), heart rate (HR), respiratory rate (RR), score on the Borg dyspnea scale (BDS), forced expiratory volume in the first second (FEV₁), forced vital capacity (FVC), forced expiratory flow of 25%–75% of FVC (FEF 25–75%), minute volume (MV), tidal volume (Vt), pulmonary rib cage volume (Vrcp), abdominal rib cage volume (Vrca) abdominal volume (Vab), inspiratory time (Ti), expiratory time (Te), total ventilatory cycle time (Ttot), duty cycle (Ti/Ttot) and frequency/tidal volume ratio (RR/Vt).

**Method**

This is an open randomized controlled crossover clinical trial. Sample selection was by convenience and patients were screened at the Pneumology outpatient facility of the Instituto de Medicina Integral Professor Fernando Figueira (IMIP) in Recife, Brazil.

The sample was calculated using a software developed by the Mallinckrodt General Clinical Research Center (MGH), based on the results of the first 10 volunteers, for a statistical power of detecting differences between procedures of 80% and a significance level of 0.05. A sample size of n = 13 patients was determined, considering that the real difference between the walk distance between procedures with and without NIV is 30 m and the standard deviation of the differences in variables 34 m.

Inclusion criteria were clinical diagnosis of CF, FEV₁ <80% of predicted, age between 7 and 16 years, clinically stable patients and with no history of hospitalization for respiratory failure in the last three months.

Exclusion criteria were patients with recent history of hemoptysis, pneumothorax or evidence of emphysematous bubbles detected by chest X-ray, Burkholderia cepacia colonization, the need for more than two physiotherapy sessions per day, gastroesophageal reflux, orthopedic trauma or cardiovascular conditions that prevents the treadmill walk test, and the presence of heart failure.
Moreover, patients with contraindication for NIV, according to the AARC GUIDELINE (2003) [21], were also excluded.

All patients agreed to take part in the study by giving their informed consent, in accordance with resolution 196/96 of the National Health Council.

Technical procedures

Patients were initially assessed for current and past conditions, medication use and anthropometric data (height, weight and body mass index — BMI). Next, a randomized plan was compiled using the Web Site Randomization.com, applying a generator of random-permuted blocks to define the order in which patients would execute the TWT, with or without NIV. A minimum and maximum rest period of 24 and 48 h, respectively, was observed between tests.

Before and five minutes after the TWT with and without NIV, patients were assessed for variations in compartmental chest wall volume and ventilatory pattern by optoelectronic plethysmography (OEP), which was followed by the spirometric test. A 30-min of NIV or rest period, at the moment when the test is done with and without NIV respectively, was given after the pulmonary function test before proceeding to the TWT.

The following procedures were involved:

**Walk test on the treadmill during 6 min:** Prior to the test, the patient was familiarized with the treadmill (G-635, Pro Action BH-fitness — Germany) and instructed about test procedures in accordance with ATS guidelines [22]. The following parameters were assessed before, immediately after and in the 5th minute post-test: blood pressure (BP), respiratory rate (RR), heart rate (HR), Borg’s scale (BS) and peripheral O2 saturation (SpO2). HR, RR and SpO2 were also monitored minute by minute during the test, such that a drop in SpO2 below 87% or rise in RR to more than 75% of age-predicted maximal HR were used as criteria for interrupting the test [23].

The test initiated with a speed of 2.5 km/h on the treadmill [24,25]. Every 30 s of the test, the examiner asked the patient if the speed could be increased, maintained or decreased, and patients answered using previously agreed upon signals. The speed could not exceed 7 km/h and if it was reached, the question would be re-asked in the next 30 s to determine whether the speed would be maintained or reduced. After the test, the walk distance (WD) on the treadmill was recorded.

The TWT performed with NIV followed the same procedures described above. However, before the test, patients were submitted to NIV on a BiLEVEL mode (Synchrony-Respironics, Brazil) for 30 min, as proposed by Serra et al. [15], with inspiratory airway pressure (IPAP) level of 12 cmH2O and final expiratory airway pressure (EPAP) of 6 cmH2O, which can vary by 2 cmH2O more or less depending on the patient’s comfort, as proposed by Holland et al. [6] and Granton and Kesten [26]. In the last six minutes of NIV the patient performed the TWT with the device.

**Optoelectronic plethysmography:** The individual was sitting with 89 retro-reflective sensors at specific thoracic and abdominal points. Eight cameras were arranged around the patient for a three-minutes recording of quiet breathing. The three-dimensional co-ordinates markers were recorded by OEP capture software (BTS Bioengineering, Italy). From this co-ordinates, the following variables were calculated by OEP before and five minutes after both tests, with and without NIV: tidal volume, minute volume, pulmonary rib cage volume, abdominal rib cage and abdominal volume of the chest wall (Vt, MV, Vrcp, Vrca, Vab), respiratory rate (RR), inspiratory time (Ti), expiratory time (Te), total ventilatory cycle time (Ttot), duty cycle, which represents the Ti/Ttot ratio and frequency/tidal volume ratio (RR/Vt).

**Spirometry:** Spirometry was conducted using a Microloop MK 8 spirometer (Micro Medical, England) to assess pulmonary function response, considering a 20-min rest period between the spirometric test and TWT. Three reproducible forced maneuvers that met American Thoracic Society criteria [27] were performed. The highest values obtained for forced expiratory volume in the first minute (FEV1), forced vital capacity (FVC), forced vital capacity...
between 25 and 75% of FVC (FEF 25–75) were recorded for analysis.

The flowchart of the methodology is represented on Fig. 1.

Statistical analysis

Statistical analysis was carried out using SPPS 20.0 statistical software, with a 95% confidence interval for all the variables.

The Shapiro–Wilk test was applied to verify normality and Levene's test for homogeneity of variances.

The study variables are presented as mean ± standard deviation, when distribution is normal, and as median and interquartile range for non-normal distribution. Inter-group comparisons were made for characterization variables using the student's t-test for independent samples.

The primary outcome, walk distance (WD) in the TWT, was analyzed by the student’s t-test for independent samples. Intra- and inter-test comparisons were performed using the Wilcoxon and Mann–Whitney tests, respectively, for cardiorespiratory (RR, HR and SpO2), and pulmonary function variables (FEV1, FVC, FEF 25–75) as well as those resulting from OEP analysis (Vt, MV, Vrcp, Vrca, Vab, Ti, Te, Ttot, Duty Cycle), measured before and after the TWT.

Results

Of the 38 eligible patients, 21 could not be contacted, three did not meet inclusion criteria and one refused to take part of the study. Seven of the 13 volunteers, 5 boys and 2 girls, started the test without NIV and after a rest period of 24–48 h did the same test with NIV. Six of 13 volunteers, 3 boys and 3 girls did the inverse order (Fig. 2).

The individual clinical characteristics are exhibited in Table 1.

All the patients reported regular medical follow-up, pulmonary impairment (mild, moderate or severe degree), and pancreatic failure with the need for enzyme and

<p>| Table 1 Basal clinical characteristics of cystic fibrosis patients studied. |
|--------------------------------|---------|----------------|----------------|</p>
<table>
<thead>
<tr>
<th>Patient</th>
<th>FEV1 (%)</th>
<th>Age (years)</th>
<th>BMI (kg/cm²)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>50</td>
<td>10</td>
<td>14.6</td>
</tr>
<tr>
<td>2</td>
<td>74</td>
<td>9</td>
<td>16</td>
</tr>
<tr>
<td>3</td>
<td>79</td>
<td>7</td>
<td>16.2</td>
</tr>
<tr>
<td>4</td>
<td>40</td>
<td>10</td>
<td>23.1</td>
</tr>
<tr>
<td>5</td>
<td>51</td>
<td>7</td>
<td>13.6</td>
</tr>
<tr>
<td>6</td>
<td>35</td>
<td>14</td>
<td>17.4</td>
</tr>
<tr>
<td>7</td>
<td>78</td>
<td>10</td>
<td>16.4</td>
</tr>
<tr>
<td>8</td>
<td>63</td>
<td>7</td>
<td>13.3</td>
</tr>
<tr>
<td>9</td>
<td>79</td>
<td>14</td>
<td>18</td>
</tr>
<tr>
<td>10</td>
<td>73</td>
<td>15</td>
<td>15.4</td>
</tr>
<tr>
<td>11</td>
<td>80</td>
<td>11</td>
<td>16.6</td>
</tr>
<tr>
<td>12</td>
<td>63</td>
<td>11</td>
<td>13.9</td>
</tr>
<tr>
<td>13</td>
<td>41</td>
<td>15</td>
<td>17.5</td>
</tr>
</tbody>
</table>

FEV1: forced expiratory volume in the first second, kg: kilogram, cm: centimeters, BMI: body mass index, M: mean, SD: standard deviation.
vitamin supplementation. Seven of the 13 patients had normal BMI for age, according to the World Health Organization (2006) \[28\], while the rest were classified as low weight. Furthermore, except for one individual, all the patients used dornase alfa.

The walk distance was greater on the TWT with NIV, with a mean of 415.38 $\pm$ 77.52 m compared to 386.92 $\pm$ 84.89 m for the same test without NIV ($p = 0.039$; Fig. 3).

Fig. 4 shows the RR, counted by ventilator pattern, HR and SpO$_2$ before, immediately after and 5 min after each test, separately and between them. Likewise, SBP, DBP and Borg’s dyspnea scale modified (BS) were also evaluated. The BS is described on Fig. 5.

With respect to the responses of cardiorespiratory variables to the TWT, there was no significant difference between the groups immediately after the tests with and without NIV. Both groups increased HR and RR, due to the rise in cardiorespiratory demand caused by physical exercise. However, only in those who underwent the TWT without NIV was there significant desaturation ($p = 0.018$), with permanent increase in RR after 5 min ($p = 0.001$). It is important to note that at no time the patients reach age-predicted HRmax. Moreover, there was no variation in intra- and inter-test SBP and DBP.

There was no inter-test difference in perceived exertion, as quantified by Borg’s scale. An increase in median and interquartile range (IR) scores occurred in both groups, immediately after the test. The post-test without NIV exhibited an increase from 0 (0–0.75) to 3 (2.5–7.5) points and in the post-test with NIV from 0 (0–0.5) to 4 (2–6) points, both with $p < 0.001$. After 5 min, median (IR) of the BS returned to 0 (0–1.5) and 1 (0–2) points on the TWT without and with NIV, respectively, significantly higher than the initial value only on the TWT with NIV ($p = 0.033$). No significant difference was found between the tests.

The spirometric variables, before and after each test, is described in Table 2. There was an increase in FEV$_1$ post-test only when exercise was performed with NIV ($p = 0.036$). There were no inter-group differences.

Fig. 6 and Table 3 show the rib cage volumes variation, as well as variables that reflect ventilatory pattern (MV, RR, Ti, Te, Ttot, Duty Cycle and RR/Vt). The total volume of the rib cage increased after exercise, but the rise was only significant when exercise was associated to NIV. Moreover, when the TWT is associated to NIV, Vt increased ($p = 0.005$), causing an alteration in thoracoabdominal kinematics, raising Vrcp and decreasing Vab ($p = 0.011$ and $p = 0.013$) respectively. There was no significant change in final values between procedures.

The TWT with NIV increased MV when compared to baseline ($p = 0.013$), increase Vt ($p = 0.005$), but did not influence the final values of RR, RR/Vt and ventilatory cycle times. There was no significant change in the ventilatory pattern in the post-test without NIV.

**Discussion**

Earlier studies in patients with CF showed NIV benefits in recruiting collapsed alveolar units, improving alveolar ventilation and oxygenation, reducing respiratory work and air trapping \[15,17,29–31\]. The present study shows that these benefits remain in a situation of cardiorespiratory overload, such as physical exercise. The previous and associated use of NIV prevented exercise-induced desaturation and reduced the time required to restore ventilatory supply-demand equilibrium after the stress test, since only on the TWT without NIV did RR remain elevated in the
5 min post-test. These benefits associated to immediate improvement in pulmonary function, observed by the increase in FEV₁ post-test, makes NIV an instrument that restores or strengthens the capacity of patients with CF to engage in physical activity.

In addition to enhancing pulmonary function and peripheral oxygenation, our findings show that the use of NIV during the TWT caused an increase in post-test Vt and MV in children and adolescents with CF.

This assessment of changes in pulmonary volumes was performed using OEP. In addition to evaluate the changes in the distribution of rib cage volume by compartments, this instrument is capable of evaluating which type of kinematic alteration is responsible for variations in Vt [32].

The heterogeneity of pleural pressure on the costal surface and the different arrangement of inspiratory and expiratory musculature in the rib cage cause different behaviors on pulmonary and thoracic mechanics in certain situations. It is therefore important to separately assess the mechanics of the rib cage in relation to the lungs (RCp) and against the diaphragm (RCd) and abdomen (RCa) [33].

We observed that exercise with NIV altered thoracoabdominal kinematics, significantly increasing Vrcp and reducing Vab. The greater expansion of the pulmonary rib cage in detriment to the reduction of Vab demonstrates that there was a difference in pressures acting on the rib cage. Cala et al. [34] reports that the increase in Vt followed by this thoracoabdominal pattern occurs due to greater recruitment of inspiratory than expiratory reserve volume [35,36,37]. Thus, the mixed pattern of restrictive and obstructive pulmonary impairment in CF was modified by the use of NIV such that Vt increased as a result of greater rib cage mobilization and a rise in its volume-generating capacity.

MV grew after exercise associated to the use of NIV due to increased Vt and not RR. So, instead of no statistically difference on RR/Vt ratio, the use of NIV tends to minimize the rapid and shallow breathing pattern after the test, suggesting an improvement in alveolar ventilation [38].

There was no difference in ventilatory cycle variables as measured by OEP. However, the basal duty cycle of cystic fibrosis patients is above normal values (between 30 and 40%). Duty cycle is the Ti/Ttot ratio of the ventilatory cycle and reflects the part of the effective diaphragm contraction cycle. High values, such as those observed in our sample, show a pattern of tachypnea, a decline in exhalatory time and greater disposition to a rise in respiratory work and respiratory muscle fatigue in situations of increased cardiorespiratory demand, such as physical exercise [39]. Moreover, mean duty cycle values were higher after exercise without NIV when compared to post-exercise values with NIV, but without statistical significance.

The present study shows that patients submitted to the TWT with NIV obtained better results, underscoring the longer walk distance and consequent enhanced functional capacity.

### Table 2 Analysis intra- and inter-test of spirometric variables, with and without NIV.

<table>
<thead>
<tr>
<th></th>
<th>TWT without NIV (n = 13)</th>
<th>TWT with NIV (n = 13)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Median (IR)</td>
<td>Intra-test p-valuea</td>
</tr>
<tr>
<td>FEV₁ (ml) (%)</td>
<td>Before 1.03 (0.83–1.7)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 1.01 (0.85–1.68)</td>
<td>0.906</td>
</tr>
<tr>
<td></td>
<td>Before 63 (40.5–76.5)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 61 (40.5–76)</td>
<td>0.608</td>
</tr>
<tr>
<td>FVC (ml) (%)</td>
<td>Before 1.75 (1.31–2.31)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 1.74 (1.23–2.24)</td>
<td>0.834</td>
</tr>
<tr>
<td></td>
<td>Before 78 (64.5–94)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 74 (59.5–94)</td>
<td>0.844</td>
</tr>
<tr>
<td>FEF 25–75 (L/s)</td>
<td>Before 0.74 (0.43–1.87)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 0.67 (0.47–1.83)</td>
<td>0.834</td>
</tr>
<tr>
<td></td>
<td>Before 38 (18–85)</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>After 36 (23.5–90)</td>
<td>0.583</td>
</tr>
</tbody>
</table>


a Wilcoxon test.
b Mann–Whitney test.
The multisystem etiology of CF and the complex physiopathology of pulmonary impairment highlight the importance of assessing the impact of NIV not only in pulmonary function, but also in functional capacity. The ability to engage in physical exercise, when objectively determined, is a more effective marker of the prognosis and quality of life in this population, which can be improved with a rehabilitation program even when the pulmonary impairment of the disease cannot be altered [29,30].

There was a significant increase in walk distance on the TWT when it was performed with ventilatory assistance. Our findings suggest that improved peripheral oxygenation and pulmonary function, caused by the use of NIV at two pressure levels, reduced the overload imposed on ventilatory muscles and increases the functional capacity of patients with CF. Accordingly, the use of NIV proved to be a necessary means of ventilatory support to supply the increased ventilatory demand and increase the functional capacity of this group of patients [12,14].

Similar results were found in patients with cardiopathies and chronic obstructive pulmonary disease, but are new in patients with cystic fibrosis in any age group [40,41].

A limiting factor of our study was the inability to assess the impact of NIV on ventilatory pattern, as measured by OEP during the test. This is due to the positive thoracic pressure exerted by NIV in cardiopulmonary interaction, modifying hemodynamics and blood volume in the chest and abdomen of individuals. These alterations in blood volume likely interfere in the assessment of total and rib cage compartment volumes [42].

Furthermore, Borg’s scale was difficult for the children to understand, limiting the validity of our results with respect to this variable. The young age and poor schooling level of the population, as well as the low socioeconomic

<table>
<thead>
<tr>
<th>Table 3</th>
<th>Analysis of volumes and ventilatory pattern by intra- and inter-test OEP.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>TWT without NIV (n = 13)</td>
</tr>
<tr>
<td></td>
<td>Median (IR)</td>
</tr>
<tr>
<td>Vt (L)</td>
<td>Before 0.31 (0.22–0.42)</td>
</tr>
<tr>
<td></td>
<td>After 0.33 (0.3–0.37)</td>
</tr>
<tr>
<td></td>
<td>After 8.185 (6.667–8.945)</td>
</tr>
<tr>
<td>RR (ipm)</td>
<td>Before 23.8 (19.4–31.8)</td>
</tr>
<tr>
<td></td>
<td>After 25.3 (20.2–28.6)</td>
</tr>
<tr>
<td>Ttot (s)</td>
<td>Before 2.5 (1.9–3.1)</td>
</tr>
<tr>
<td></td>
<td>After 2.4 (2.1–3)</td>
</tr>
<tr>
<td>Ti (s)</td>
<td>Before 1.06 (0.85–1.47)</td>
</tr>
<tr>
<td></td>
<td>After 0.99 (0.85–1.39)</td>
</tr>
<tr>
<td>Te (s)</td>
<td>Before 1.47 (1.05–1.95)</td>
</tr>
<tr>
<td></td>
<td>After 1.38 (1.2–1.69)</td>
</tr>
<tr>
<td>Duty cycle (%)</td>
<td>Before 43.1 (40.2–47.2)</td>
</tr>
<tr>
<td></td>
<td>After 43.3 (38.4–45.2)</td>
</tr>
<tr>
<td>RR/Vt (ipm/L)</td>
<td>Before 80.0 (44.4–101.4)</td>
</tr>
<tr>
<td></td>
<td>After 75.2 (57.8–98.5)</td>
</tr>
</tbody>
</table>


a Wilcoxon test.

b Mann–Whitney test.
status of the sample may have been the cause of this limitation. Hook et al. [43] showed that low economic status has an influence in a wide range of situations, including cognitive skills, school performance, executive ability and even physical and mental health.

Conclusion

The pulmonary impairment in cystic fibrosis patients can increase the ventilatory demand even in performing their activities of daily living. Dyspnea being an important limiting factor in their functionality and quality of life.

Given the impossibility of altering the severity of pulmonary impairment, the present study proved that the immediate use of NIV in the BiLEVEL mode improves pulmonary function, respiratory muscle strength, peripheral oxygenation, as well as providing the ventilatory support required to increase the functional capacity of children and adolescents with CF.

Studies that assess the impact of NIV associated to a pulmonary rehabilitation program on long-term functional capacity, health gains, physical conditioning and functionality scale should be encouraged.

Conflict of interest

None declared.

References

[21] AARC guideline: intermittent positive pressure breathing. Respir Care 2003;48(5).


