Home mechanical ventilatory support in patients with restrictive ventilatory disorders: A 48-year experience

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Summary We performed a retrospective analysis to the effects of negative pressure ventilation (NPV), tracheal intermittent positive pressure ventilation (TIPPV), and nasal intermittent positive pressure ventilation (NIPPV, volume or pressure-controlled ventilatory mode), in 114 patients with restrictive ventilatory disorders instituted in our hospital from 1956 until 2005. The patients were assigned on “ad hoc” basis to NPV, TIPPV, or NIPPV.

All patients were subdivided in an idiopathic kyphoscoliosis group (IK, n = 64), a post-polioomyelitis syndrome group (PP, n = 30), or a miscellaneous group (M, n = 20). The patients in the PP group had higher survival rates compared to the IK patients (P < 0.05), while the M patients had the lowest survival rates (P < 0.01). Both NPV (P < 0.01) and TIPPV (P < 0.05) lead to a decrease in PaCO₂ after 9 months compared to baseline. This decrease in PaCO₂ was still present after 5 years NPV (P < 0.001) and TIPPV (P < 0.05). NIPPV lead to an improvement in pulmonary function (P < 0.05) and arterial blood gases (P < 0.001) after 9 months compared to baseline. After 5 years NIPPV, the arterial blood gases were still significantly improved compared to baseline (P < 0.01). Both volume-controlled and pressure-controlled ventilation improved pulmonary function and arterial blood gases.

Conclusion: Long-term home mechanical ventilatory support by both NPV and positive pressure ventilation is effective in patients with IK, PP syndrome, and a M group, even after a period of 5 years.
Introduction

Restrictive ventilatory disorders are characterized by a reduced chest wall compliance and mechanical disadvantage of the respiratory muscles leading to a decrease in respiratory function and an increase in the work of breathing. Therefore, patients with restrictive ventilatory disorders will adopt a pattern of rapid, shallow breathing, which may result in chronic alveolar hypoventilation. During sleep, there is a progressive fall in minute ventilation exaggerating the hypoventilation. As a consequence, these patients develop hypercapnia, firstly during sleep, and finally in wakefulness. Eventually, respiratory failure may occur, which makes chronic ventilatory support necessary.

Home mechanical ventilation (HMV) has been used for more than 60 years now in a variety of disorders. Several methods have been used: firstly only negative pressure ventilation (NPV) was available, later on tracheal intermittent positive pressure ventilation (TIPPV) appeared, and finally nasal intermittent positive pressure ventilation (NIPPV) became available.

NPV devices are cumbersome to use and may lead to insufficient ventilation due to an inadequate covering of the chest wall surface and the abdomen. In our hospital we solved this problem by using a specially designed tailor-made shell. Despite the disadvantages of NPV, it has shown beneficial effects in patients with a variety of pulmonary disorders, including patients with restrictive ventilatory disorders.

TIPPV has been shown to be effective in patients with restrictive ventilatory disorders. Although the use of TIPPV is limited by drawbacks such as disfigurement, difficulties associated with phonation, risk of infection, and the burden of tracheostomy care, this therapy is nowadays still prescribed to obtain adequate ventilatory support.

NIPPV is nowadays the most frequently used mode of ventilatory support in patients with restrictive ventilatory disorders. Several studies have shown the benefits of NIPPV in terms of improvement in daytime arterial blood gas tensions, relief of nocturnal hypoventilation and its symptoms, improvement of health-related quality of life, and improved survival. At the introduction of NIPPV, only volume-controlled ventilators were available. Later, pressure-controlled and bilevel pressure-controlled ventilation became available as well. Only a few studies have compared the effects of volume-controlled vs. pressure-controlled ventilation.

However, a number of issues are remarkable from the above-mentioned studies. Firstly, many of them only monitored the effects of non-invasive ventilatory support, while invasive ventilation is still being used in a considerable number of patients. Secondly, most studies assessed the effect of ventilatory support for a short period. Finally, patients with a variety of different disorders were placed and analyzed in the same group to increase the number of patients in the study groups.

In our hospital we have been using HMV since 1956 in a variety of disorders. For this study, we selected the patients with kyphoscoliosis, post-polioymyelitis (PP) syndrome, and other restrictive ventilatory disorders for analysis of the effects of HMV, as we have been able to build up long-term experience with HMV in a large number of patients with restrictive ventilatory disorders.

Therefore, the aim of the study was: (a) to describe the development of HMV in our hospital, and (b) to assess the effects of NPV, TIPPV, and NIPPV on pulmonary function, arterial blood gas tensions, and survival and in patients with kyphoscoliosis, post-polioymyelitis syndrome and miscellaneous restrictive ventilatory disorders. Therefore we set up a retrospective analysis including all 114 patients with restrictive ventilatory disorders who received ventilatory support in our hospital from 1956 until 2005.

Methods

Subjects

Data were collected from all patients who received HMV initiated at our department of HMV from 1956 until 2005. From this population we selected patients with idiopathic kyphoscoliosis (IK), PP syndrome and other restrictive ventilatory disorders for analysis of the effects of HMV.

Ventilatory equipment and monitoring

Patients were ventilated by NPV, TIPPV, or NIPPV. NPV was delivered by means of a chest respirator with tailor-made shell. TIPPV was delivered by means of volume or pressure ventilation. NIPPV was delivered via nasal or full face mask, either by a volume-controlled ventilator (Monnal D, Taema, Antony Cedex, France; Lifecare PLV 100, Respironics, Murrysville, USA; Breas PV 501, Breas Medical, Mölndal, Sweden), or a pressure-controlled ventilator (bilevel pressure (BilevelPAP; Puritan Bennett PB 335, Respironics, Murrysville, USA; Breas 401, Breas Medical, Mölndal, Sweden); or pressure-controlled (PCV; Airox VP 2000, Beaumont, Migennes, France; Breas 401, Breas Medical, Mölndal, Sweden).
Although nowadays we have the choice between volume-controlled or pressure-controlled ventilators, in the past the decision to implement a particular mode of ventilation was dependent on the availability of the ventilator in the market at the moment patients were instituted.

All patients were instituted on ventilatory support in the hospital. The indication for chronic ventilatory support was either chronic stable or progressively deteriorating respiratory failure unresponsive to other treatment options, both in combination with symptoms such as increasing shortness of breath on exertion, tiredness, and sleepiness.

Ventilator settings were determined at baseline and adjusted depending on arterial blood gas tensions. Before 1990, we measured arterial blood gas tensions at rest during the day. Since 1990, we performed an arterial blood gas registration in the hospital during the night, initially without the ventilator, and after the patients were able to tolerate the ventilation for at least 6 h a night, with the ventilator. Thereafter they were discharged and followed at the outpatient clinic. After 2 months we performed another overnight arterial blood gas registration while on the ventilator and adjusted ventilator settings as necessary. Furthermore, the patients were monitored every 6 months, including pulmonary function tests, daytime arterial blood gas levels (ABG), and end-tidal CO₂ measurements.

Supplemental oxygen was provided in patients in whom arterial oxygen saturation remained low (saturation <90%) despite optimal ventilator settings.

Data collection and analysis

We collected the following data: birth date, sex, primary, and secondary diagnoses, indication for initiating ventilatory support, date of starting and ending ventilatory support, mode of ventilatory support, oxygen need, and prescribed hours of ventilatory support. Furthermore, we collected data on pulmonary function, nocturnal and daytime arterial blood gas analysis, and dependency in activities of daily living (ADL) just before ventilatory support was initiated (baseline) and after 9 months, 1½ years, 3 years, and then every 2 years after initiating ventilatory support. Arterial blood gas tensions were obtained while the patients were breathing room air without ventilation.

Statistical analysis

Survival rates were calculated and compared between the three diagnostic groups by using the method of Kaplan–Meier and log rank tests. Differences in the age at start between the diagnostic groups (IK, PP, and M) were assessed by a Kruskal Wallis test; differences in baseline arterial blood gases and pulmonary function were assessed by one-way analysis of variance.

We compared baseline arterial blood gases and pulmonary function with the values obtained after 9 months (short-term effects) and to those after 5 years (the long-term effects) by multiple linear regression of repeated measurements. Patients who dropped out and patients from whom only incomplete data could be collected were excluded from these analyses.

Differences in baseline parameters between patients ventilated by volume-controlled and pressure-controlled NIPPV were assessed by Wilcoxon sign rank tests. In the patients receiving volume-controlled and pressure-controlled ventilation, we compared baseline arterial blood gases and pulmonary function with the values obtained after 9 months by Mann–Whitney U tests.

Results

HMV at the University Medical Center Groningen

From 1956 until January 1, 2005 we instituted 433 patients with a wide variety of disorders on HMV at our hospital. We instituted patients with restrictive ventilatory disorders (IK, PP complicated by kyphoscoliosis, post-tuberculosis); pure neuromuscular disorders (morbus Duchenne, amylotropic lateral sclerosis); pulmonary disorders (COPD patients, cystic fibrosis patients); and patients with an obstructive sleep apnea syndrome (OSAS) or obesity hypoventilation syndrome (OHS). The treatment prevalence of HMV in region of the HMV Center Groningen was 5/100.000 on January 1, 2000 and 8.5/100.000 on January 1, 2005. This increase is mainly caused by a more than fivefold increase in the number of patients with ALS (2000: 6 active users; 2005: 41 active users) and a more than twofold increase in the number of patients with OSAS or OHS (2000: 20; 2005: 44) (Fig. 1).

Characterization of the study subjects

We selected the patients with IK, the PP syndrome, and other restrictive ventilatory disorders for analysis of the effects of HMV. We subdivided these patients in three groups according to their primary diagnosis. The first group included 64 patients (25
men, 39 women) with IK. The second group included 30 patients (15 men, 15 women) with the PP syndrome. A third group included 20 patients with a miscellaneous restrictive ventilatory disorder. This group consisted of 12 patients (3 men, 9 women) who underwent a thoracoplasty and/or (partial) lung resection for tuberculosis (performed between 1939 and 1953), 5 patients (3 men, 2 women) who experienced spondylitis tuberculosa, 2 women with bronchiectasis (one of them underwent a thoracoplasty in 1939), and 1 woman with atelectase as a result of radiotherapy for lung metastases. This group was called the miscellaneous (M) group. Baseline characteristics of the three diagnostic groups are shown in Table 1.

Table 1  Number of patients and baseline characteristics all patients per diagnostic group.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number (n)</th>
<th>Proportion of acute patients (%)</th>
<th>Age at start (years)</th>
<th>FEV1 (L)</th>
<th>VC (L)</th>
<th>PaO2 (kPa)</th>
<th>PaCO2 (kPa)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IK</td>
<td>64</td>
<td>39</td>
<td>56 (13.4)</td>
<td>0.77 (0.27)</td>
<td>1.10 (0.44)</td>
<td>7.44 (2.04)</td>
<td>7.77 (1.72)</td>
</tr>
<tr>
<td>PP</td>
<td>30</td>
<td>43</td>
<td>50 (15.8)</td>
<td>0.76 (0.35)</td>
<td>1.16 (0.61)</td>
<td>8.33 (1.56)</td>
<td>7.33 (1.34)</td>
</tr>
<tr>
<td>M</td>
<td>20</td>
<td>50*</td>
<td>64† (10.2)</td>
<td>0.64 (0.15)</td>
<td>1.09 (0.43)</td>
<td>8.88† (1.81)</td>
<td>8.01 (2.19)</td>
</tr>
</tbody>
</table>

Values are expressed as mean (SD).
Note: VC: vital capacity; FEV1: forced expiratory volume in 1 s; PaO2: arterial oxygen pressure; PaCO2: arterial carbon dioxide pressure.
* M higher than IK (P<0.001).
† M higher than IK (P<0.05) and PP (P<0.01).
‡ M higher than IK (P<0.05).

A total of 48 patients started ventilatory support because of acute on chronic respiratory failure (acute patients). For the other patients, ventilatory support was initiated non-emergently because of chronic ventilatory failure with symptoms like shortness of breath on exertion, tiredness, sleepiness, and morning headache (chronic patients). The proportion of patients instituted acutely was higher in the M group than in the IK group. At baseline, the chronic patients had a significantly higher vital capacity (VC; \( P<0.05 \)), higher daytime \( \text{PaO}_2 \) (\( P<0.003 \)), and lower daytime \( \text{PaCO}_2 \) (\( P<0.001 \)) compared to the acute patients.

Survival and causes of death in three different groups of patients

Figure 2 shows the cumulative survival of patients from the three diagnostic groups receiving ventilatory support. The PP patients had higher survival rates than the IK patients (\( P<0.05 \)). The M patients group experienced the lowest survival rates (\( P<0.01 \)) of all groups.

Of the 64 IK patients, 40 patients were still being ventilated at our hospital at the time of the analysis with a median duration 4.5 years
Twenty IK patients had died after a median duration of 6.3 years HMV (interquartile range 6.6); 11 patients died from respiratory failure, 1 patient died from esophageal cancer, 1 patient died from a malignant sarcoma, 1 patient got a fatal accident, and 6 patients died from unknown causes. Furthermore, 3 IK patients were lost from follow-up and 1 patient decided to end the HMV after 1 month because of coping problems. We found no significant differences in age at start of ventilation, baseline pulmonary function and arterial blood gas tensions between the IK patients who had died and the IK patients who were still alive at the time of the analysis. However, the IK patients who had died had more frequently been ventilated by TIPPV (25%) or NPV (30%) compared to the IK patients who were still alive at the time of the analysis (TIPPV: 7.5%, NPV: 5%).

Of the 30 PP patients, 21 patients were still being ventilated at the time of the analysis after a median duration of 11.3 years (interquartile range 11.0). Nine PP patients had died after a median duration of 10.3 years ventilatory support (interquartile range 20.8); 4 patients died from respiratory failure, 2 patients died after surgery for a hip fracture, 1 patient died from a cerebral hemorrhage, and in 2 patients died from unknown causes. We found no significant differences in age at the start of ventilation, pulmonary function, arterial blood gas tensions, or type of ventilatory support at baseline between the PP patients who had died and the PP patients who were still alive at the time of the analysis.

Of the 20 M patients, 5 patients were still being ventilated at the time of the analysis after a median duration of 5.1 years HMV (interquartile range 3.4). Eleven M patients had died after a median duration of 4.7 years HMV (interquartile range 5.5); 3 M patients died from respiratory failure, one patient died from a heart attack, 2 patients died after they became severely depressed and quitted the assisted ventilation voluntarily, and 4 patients died from unknown causes. Furthermore, 3 M patients ended the ventilatory support (2 NIPPV ventilated patients, 1 NPV ventilated patient) because of coping problems after a median duration of 14 months ventilatory support (interquartile range 40), and 1 patient was lost from follow-up. We found no significant differences in age at start of ventilation, pulmonary function, and arterial blood gas tensions at baseline between the M patients who had died, the M patients who were still alive and the M patients who had stopped because of coping problems. The M patients who were still being ventilated at the time of the analysis were all instituted non-emergently. In contrast, the M patients who had died were more frequently instituted acutely (9 patients instituted acutely, 2 patients non-emergently).

Figure 2 Cumulative survival of idiopathic kyphoscoliosis patients, post-poliomyelitis patients, and patients with miscellaneous restrictive ventilatory disorders, treated by home mechanical ventilation (HMV).
At baseline, a significantly higher proportion of IK patients (73%) compared to the PP patients (47%) and M patients (40%) was independent in ADL (P < 0.01). After 5 years HMV, significantly less IK patients were ADL independent (54%) compared to baseline. In the PP group the proportion of ADL independent patients was not changed (47%) after 5 years HMV compared to baseline. In the M group only 25% of the M patients was independent in ADL after 5 years HMV compared to baseline (not significant).

Different types of ventilatory support

**NPV**

Twenty patients received NPV (9 IK patients, 8 PP patients, 3 M patients). Sixteen patients were ventilated with NPV for at least 5 years (80%). NPV significantly improved PaCO₂ after 9 months and even after 5 years HMV (Table 2).

Only 1 IK patient and 1 PP patient changed from NPV to TIPPV after 30 and 279 months, respectively. One PP patient received 48 months cuirass followed by 32 months NIPPV before he eventually switched to TIPPV. The reason for switching to invasive ventilatory support was deterioration into respiratory failure in all three patients. We found no significant differences in baseline lung function parameters and baseline daytime or overnight arterial blood gas values between the patients who switched to TIPPV and the patients who remained receiving ventilatory support by NPV.

**TIPPV**

Sixteen patients received TIPPV (9 IK patients, 4 PP patients, 3 M patients). Eleven patients were ventilated by TIPPV for at least 5 years (69%). TIPPV improved PaCO₂ after 9 months compared to baseline, and this effect was still evident after 5 years (Table 2).

Two patients were instituted on TIPPV because of chronic respiratory failure. One of those patients got TIPPV in 1975 while he already received a tracheostoma in 1968 for reducing dead space area of the lungs, while in the other patient TIPPV was used after NIPPV failed because of severe apneas during NIPPV. Fourteen patients were instituted on TIPPV after a period of severe acute respiratory failure. This group consisted of 6 patients in whom NIPPV was tried but failed repeatedly because no improvement in gas exchange could be obtained or because of severe sputum clearance problems, 4 patients who were set on TIPPV directly after a period of intubation because of expected weaning problems, 2 patients instituted on TIPPV before NIPPV was available, and 2 patients who were already tracheostomized in another hospital.

Later on, 3 patients changed from TIPPV to NIPPV (1 patient from the IK group after 4 months (in 2000) and 2 patients from the PP group after 9 months (in 1993) and 52 months respectively (in

<table>
<thead>
<tr>
<th>Ventilatory mode</th>
<th>Baseline</th>
<th>9 months</th>
<th>5 years</th>
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<tbody>
<tr>
<td><strong>NPV</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>FEV₁ (n = 7)</td>
<td>0.78 (0.49)</td>
<td>0.96 (0.46)</td>
<td>0.93 (0.37)</td>
</tr>
<tr>
<td>VC (n = 7)</td>
<td>1.06 (0.66)</td>
<td>1.16 (0.68)</td>
<td>1.23 (0.55)</td>
</tr>
<tr>
<td>PaO₂ (n = 9)</td>
<td>6.97 (1.98)</td>
<td>8.79 (1.82)</td>
<td>8.26 (1.76)</td>
</tr>
<tr>
<td>PaCO₂ (n = 9)</td>
<td>8.58 (1.08)</td>
<td>6.54 (0.88)**</td>
<td>6.52 (0.69)**</td>
</tr>
<tr>
<td><strong>TIPPV</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>FEV₁ (n = 4)</td>
<td>0.69 (0.18)</td>
<td>0.74 (0.21)</td>
<td>0.70 (0.24)</td>
</tr>
<tr>
<td>VC (n = 5)</td>
<td>0.95 (0.36)</td>
<td>1.08 (0.32)</td>
<td>1.10 (0.43)</td>
</tr>
<tr>
<td>PaO₂ (n = 4)</td>
<td>6.89 (1.63)</td>
<td>9.14 (1.73)</td>
<td>9.36 (1.86)</td>
</tr>
<tr>
<td>PaCO₂ (n = 6)</td>
<td>9.71 (1.97)</td>
<td>5.85 (0.76)*</td>
<td>6.00 (0.53)*</td>
</tr>
<tr>
<td><strong>NIPPV</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>FEV₁ (n = 27)</td>
<td>0.81 (0.28)</td>
<td>0.91 (0.31)*</td>
<td>0.87 (0.27)</td>
</tr>
<tr>
<td>VC (n = 29)</td>
<td>1.29 (0.58)</td>
<td>1.42 (0.63)*</td>
<td>1.39 (0.64)</td>
</tr>
<tr>
<td>PaO₂ (n = 22)</td>
<td>7.86 (1.95)</td>
<td>9.60 (1.45)**</td>
<td>9.39 (1.30)**</td>
</tr>
<tr>
<td>PaCO₂ (n = 29)</td>
<td>7.46 (1.37)</td>
<td>5.96 (0.75)***</td>
<td>6.23 (0.73)***</td>
</tr>
</tbody>
</table>

Values are expressed as mean (SD).

Note: VC: vital capacity; FEV₁: forced expiratory volume in 1 s; PaO₂: arterial oxygen pressure; PaCO₂: arterial carbon dioxide pressure.

*P < 0.05 (compared to baseline); **P < 0.01 (compared to baseline); ***P < 0.001 (compared to baseline).
1999)). In 1 other patient an attempt was made to switch to NIPPV, unsuccessful due to anxiety. The other 12 patients stayed on TIPPV. Half of them died before 1986 when NIPPV was not available yet at our hospital. In the other half, because the patients had no complaints about the TIPPV, the decision was made not to change a successful treatment.

**NIPPV**

Seventy-eight patients received NIPPV (46 IK patients, 18 PP patients, 14 M patients). Thirty-four patients received NIPPV for at least 5 years (48.6%). NIPPV improved pulmonary function and ABG after 9 months compared to baseline, and the improvement in arterial blood gases was still evident after 5 years NIPPV (Table 2).

Only 1 IK patient switched from NIPPV to TIPPV after 25 months because NIPPV could not give a satisfactory relief of his clinical condition.

**Volume or pressure support**

Of the 78 patients on NIPPV, 27 patients received volume-controlled ventilation (11 IK patients, 12 PP patients, 4 M patients), 17 patients pressure-controlled ventilation (11 IK patients, 2 PP patients, 4 M patients), and 28 patients bilevelPAP (21 IK patients, 4 PP patients, 3 M patients). Data regarding the type of NIPPV were lost in 6 patients and these patients were excluded from the analyses.

Changes in home mechanical ventilatory support modes are shown in Fig. 3. The first patients who received NIPPV were instituted in 1989. In the first years mostly volume-controlled ventilation was available. As a relatively large percentage of the patients instituted on NIPPV in the first years were diagnosed with PP syndrome, most of these patients were instituted on volume-controlled ventilation. These days, there is an almost equal proportion of volume-controlled and pressure-controlled ventilation in our hospital.

The effects on pulmonary function and gas exchange are presented in Table 3. Both volume and pressure-controlled ventilation improved pulmonary function and arterial blood gas tensions.

**Daily ventilator use**

All the patients used their ventilator during the night. At baseline, several patients used the ventilator during the day as well (10 patients on NIPPV (13%), 7 patients on TIPPV (44%) and 3 patients on NPV (16%)).

After 1½ years, next to overnight use, significantly more patients on NIPPV used their ventilator during the day (34% \(P < 0.05\)). In the TIPPV ventilated patients (36%), and in the NPV ventilated patients (27%) the degree of daytime use did not change significantly.

**Discussion**

The present study illustrates that both invasive and non-invasive HMV are effective in terms of

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**Figure 3** Changes in ventilatory support mode of NIPPV from 1989 until 2005 in the University Medical Center Groningen.
short-term and long-term improvement of pulmonary function and arterial blood gas tensions in patients with kyphoscoliosis, PP syndrome, or a miscellaneous restrictive ventilatory disorder. Furthermore, the study demonstrates that volume-controlled and pressure-controlled ventilation are equally effective in improving pulmonary function and arterial blood gas tensions in patients with restrictive ventilatory disorders.

In this study, we selected the patients with restrictive ventilatory disorders to analyze the effects of HMV. Last years, the number of patients on HMV in the area covered by the department of HMV of the University Medical Center Groningen has increased. These data agree with data from Sweden. This increase in prevalence is mainly due to an increase in the number of patients with neuromuscular disorders, such as ALS, and patients with OSAS/OHS that were instituted on HMV. However, because the restrictive ventilatory disorder group has remained relatively constant over several years now, we have been able to build up long-term experience with HMV in a large number of patients.

The IK and the PP patients showed survival rates of 84% and 93% after 5 years, respectively. These survival rates are comparable to previous studies in these patients. The M group showed a significantly worse survival rate of 62% after 5 years compared to the IK and PP patients. The worse survival rate of the M patients can partly be explained by the older age at the start of HMV in these patients compared to the IK and PP patients. Furthermore, it is important to note that the M patients were mainly patients who experienced tuberculosis and therefore had an underlying intrinsic lung disease. In the study of Jackson et al., patients who underwent a thoracoplasty had a 5-year survival rate of 64%, which is comparable to the survival of our M group.

Two patients in our M group suffered from bronchiectasis. Although it has been shown that patients with bronchiectasis have very low survival rates, the three bronchiectasis patients of the M group in our study had a moderate survival rate. One patient that initiated cuirass at age 47 died after 16 years of ventilatory support, the other patient that initiated NIPPV at age 70 died after 9 years ventilatory support. The long survival of these patients tells us that not all patients with bronchiectasis are bad candidates for chronic ventilatory support.

**NPV**

Several studies have shown benefits from long-term home NPV in patients with restrictive ventilatory disorders. However, direct comparisons between NPV and NIPPV are rare. Baydur et al. described the outcomes of 79 patients receiving home ventilation by NPV or NIPPV. They found that 25 patients with poliomyelitis VC and PaCO2 did not change significantly on body ventilation (tank or shell) and that NIPPV resulted in better outcomes in terms of a decreased number of tracheotomies and a better survival rate compared to NPV. We did find an improvement of pulmonary function and ABG and, although in the IK group there seemed to be a survival benefit for the NIPPV ventilated patients, we did not find a difference in overall survival rates between the patients on NPV and those on NIPPV. Furthermore, in our study only 2 patients on NPV switched to TIPPV (10%) compared to 56% in the study of Baydur.

However, patient characteristics and diagnoses of the patients were different in the study of Baydur and in our study. Secondly, as more than half of Baydur’s patients were instituted on ventilatory support during the primary phase of poliomyelitis at a young age, the duration of NPV in the study of Baydur (mean 24 years) was far longer than in our study (mean 12.5 years). This might explain the high number of tracheotomies in the study of Baydur, compared to our study.

On the basis of outcomes in terms of physical effects, no conclusion can be drawn about superiority
of NPV or NIPPV. However, because of the difficult and cumbersome use of NPV, nowadays NIPPV tends to be the first choice. Our study again shows that NPV can be effective in restrictive ventilatory disorders. This supports the idea that NPV remains a second choice to be used in patients whom, for technical or other reasons, cannot be offered NIPPV.36

TIPPV

In our study 20 patients started with TIPPV. Most of our patients on TIPPV required this type of ventilation because of acute on chronic respiratory failure which could not be controlled by NIPPV or because of uncontrollable sputum clearance problems. However, with the availability of a variety of nasal and mouth interfaces, coughing techniques and machines, it became increasingly possible to use NIPPV instead of TIPPV, even in acute situations. In the period from 1997–2005, only 1 PP patient eventually required chronic TIPPV, because of deteriorating blood gasses and a deteriorating clinical condition on NIPPV.

Three of the patients on TIPPV changed successfully to NIPPV. In two other patients, severe sputum clearance problems and anxiety for not being adequately ventilated at night hindered a successful switch to TIPPV. The patients required TIPPV in the acute situation but were in a stable condition at the time of the switch. In the future, we should try to switch more tracheostomized patients to TIPPV, because of deteriorating blood gasses and a deteriorating clinical condition on NIPPV.

Our results agree with the study of Zaccaria et al. who found that in patients with respiratory insufficiency who were treated by TIPPV arterial blood gasses improved to a same degree than in patients treated by NIPPV. Furthermore, they found that these effects were still evident after 1 year of ventilation.10

NIPPV

Several studies have shown that NIPPV is effective in patients with restrictive ventilatory disorders.11–28 In our study, NIPPV improved ABG and pulmonary function, and these positive effects were significant even after of 5 years ventilatory support. Only 1 patient with chronic hypoxia, pulmonary hypertension, and heart failure changed to TIPPV after 25 months of NIPPV because severe air leakage hindered an adequate oxygenation.

Several potential mechanisms are postulated to explain the effects of NIPPV. It may improve the mechanical properties of the thorax, it may “rest” the respiratory muscles, and it may improve respiratory drive.31,37 We can only suggest that, in our patients, NIPPV worked through improving the mechanical properties of the thorax (as we did find a small improvement in VC and FEV1), probably in combination with the other mechanism mentioned above. However, we did not measure respiratory muscle strength, respiratory muscle activity, or CO2 sensitivity.

According to the Conference Consensus of 1999, daytime hypercapnia in combination with symptoms of shortness of breath on exertion, tiredness, sleepiness, and ankle swelling is the primary indication for starting NIPPV.38 Furthermore, we found that the success of HMV seems to be linked to the clinical condition of patients at the time they were instituted. The patients who were instituted acutely had worse baseline arterial blood gasses and pulmonary function, and seemed to have a worse survival rate on HMV than the patients instituted non-emergently. Furthermore, more patients suffering from acute respiratory failure had to be trachestomized. Therefore, it seems better to initiate ventilator support before patients deteriorate into acute respiratory failure. Recently it has been suggested that starting NIPPV at the stage of nocturnal hypoventilation before daytime hypercapnia ensues can prevent ventilatory decompensation,39 which suggests that it is a good policy to initiate NIPPV even earlier in the course of the disease, maybe even before daytime hypercapnia develops.

In summary, NPV, TIPPV, and NIPPV have a positive effect on pulmonary function and arterial blood gas tensions both at short-term and long-term. If patients receive NIPPV the mode of ventilation did not show different effects on pulmonary function and ABG. Patients with IK and PP syndrome showed a significantly better survival rate compared to the M group. As the underlying disorder determines survival and probably the course of the disease, we think it is important to assess the effects of HMV in patients with different disorders separately.

References


