# Home Treatment of Infection-Related Acute Respiratory Failure in Kyphoscoliotic Patients on Long-Term Mechanical Ventilation

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BACKGROUND: In patients with kyphoscoliosis, long-term mechanical ventilation improves chronic alveolar hypoventilation during spontaneous breathing, improves quality of life, decreases the need for hospitalization, and improves survival. In these patients respiratory infection can precipitate acute respiratory failure (ARF) that requires hospitalization. OBJECTIVE: To study the possibility of home treatment of infection-related ARF in kyphoscoliotic patients on long-term mechanical ventilation. METHODS: During a period of 4 years, 8 kyphoscoliotic patients (3 women, 5 men, mean  $\pm$  SD age 61  $\pm$  10 y, mean Cobb angle 84  $\pm$  7°), who had been using overnight mechanical ventilation (delivered by either volume-limited [4] or pressure-limited [4] ventilators) for  $31 \pm 32$  months, developed infection-related ARF. Seven patients agreed to be treated at home, with an increase of the daily duration of mechanical ventilation to > 20 hours, and antibiotics. Blood oxygen saturation was monitored via pulse oximetry during mechanical ventilation and overnight, to determine whether to add or increase supplemental oxygen. A nurse, a general practitioner, and a chest specialist made scheduled visits to each patient. RESULTS: All 7 patients were successfully treated at home. In 2 patients supplemental oxygen flow was slightly increased. Two patients who had not previously been receiving supplemental oxygen received supplemental oxygen for a few days. The patients progressively decreased the daily duration of mechanical ventilation, according to their ability to breathe comfortably without mechanical assistance, under the supervision of the medical staff, and they all returned to their baseline (pre-ARF) condition in 4 weeks. CONCLU-SION: In kyphoscoliotic patients on long-term mechanical ventilation, home treatment of infectionrelated ARF is possible and effective, provided there is adequate collaboration by the patients and their relatives, and staff well-trained in mechanical ventilation and other aspects of the home care of these patients. Key words: mechanical ventilation, home care, kyphoscoliosis, acute respiratory failure, noninvasive ventilation. [Respir Care 2007;52(6):713-719. © 2007 Daedalus Enterprises]

#### Introduction

In patients with kyphoscoliosis, impaired ventilatory mechanics may cause chronic alveolar hypoventilation and

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ventilatory insufficiency.<sup>1–3</sup> Long-term mechanical ventilation, when used 8–12 h/d, improves arterial blood gas values during spontaneous breathing, enhances quality of life, reduces the hospitalization rate, and increases surviv-

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al.<sup>4–12</sup> Respiratory tract infections can rapidly worsen these patients' respiratory condition and precipitate acute respiratory failure (ARF), which usually requires hospitaliza-

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tion and either noninvasive positive-pressure ventilation (NPPV) or intubation and mechanical ventilation. In such cases the conventional treatment is to increase the daily duration of mechanical ventilation and to treat the infection with antibiotics. We prospectively studied the hypothesis that home care of such cases can be as safe and effective as hospital care.

#### Methods

Our program of long-term mechanical ventilation started in 1993. Patients and their families are trained to use mechanical ventilators and are visited at home once a month (or on request from the specialized nursing staff) to check the patient's condition and ensure correct ventilator use. The patients also have a yearly hospital visit that includes pulmonary function tests (MasterLab, Jaeger, Friedberg, Germany), arterial blood gas (ABG) testing (AVL Omni 6, Diamond Diagnostics, Holliston, Massachusetts), and overnight pulse oximetry (Nonin 8500, Life Assist, Rancho Cordova, California), and ventilator settings are adjusted as necessary.

During a 4-year period (1998–2002), we prospectively studied our patients with idiopathic severe kyphoscoliosis (Cobb angle  $> 70^{\circ}$ ), who had been on mechanical ventilation for at least 6 months, and who called their general practitioner because of symptoms of respiratory infection.

The inclusion criteria were tachypnea (> 30 breaths/min), cyanosis, purulent secretions, cough, and fever. In such cases the chest specialist, the general practitioner, and the nurse of the long-term mechanical ventilation service together evaluated the patient at the patient's home. Arterial blood samples were obtained during spontaneous breathing and processed immediately at the patient's home, with a portable analyzer (StatPal I, PPG Industries). Patients with respiratory acidosis (pH 7.35-7.25) were mechanically ventilated and after 1 hour another arterial blood sample was analyzed. We also collected electrocardiogram readings and expectorated sputum samples for bacterial identification. Supplemental oxygen was administered as needed during mechanical ventilation to keep their oxygen saturation (measured via pulse oximetry  $[S_{pO_2}]$ ) over 90%. Mechanical ventilation was administered continuously until the morning. S<sub>pO2</sub> was recorded continuously all night. If the patient showed improvement in ABG values 1 hour after initiating mechanical ventilation and his or her mean nocturnal  $S_{pO_2}$  was > 90%, the patient was invited to participate in the home treatment protocol.

This study was performed in accordance with the Helsinki Declaration. Informed consent was obtained from the patients enrolled in the study. Each patient gave written informed consent to be treated at home rather than in hospital.

The exclusion criteria were chronic obstructive pulmonary disease (COPD) (ratio of forced expiratory volume in the first second to forced vital capacity < 70%), bronchial asthma, symptomatic cardiovascular disease, substantial cardiac arrhythmia, and abnormalities of consciousness (Glasgow Coma Score < 14).

Included patients were asked to use nearly continuous mechanical ventilation (> 20 h/d) for the first week, with the ventilator settings they used in their baseline condition, and to progressively decrease the daily duration of mechanical ventilation according to the tolerance of spontaneous breathing, under the supervision of the medical staff. All the patients were treated with antibiotics for 2 weeks (usually oral clarithromycin, 500 mg, twice a day, and intramuscular ceftazidime, 1 g, 3 times a day) and with metered-dose-inhaler Albuterol, 4 times a day, via holding chamber (Aerosol Cloud Enhancer, DHD Healthcare Wampsville, New York) during mechanical ventilation. Once a week, arterial blood samples were collected after 1 hour of spontaneous breathing without the ventilator. Sputum collection and culture was repeated 2 weeks after starting the antibiotics.

During the study period the nurse visited the patient's home 3 times a day. The first visit lasted 1 hour and each subsequent nurse visit lasted about 20 min. During each visit, the nurse checked the ventilator's hour counter, the  $S_{pO_2}$  during spontaneous breathing and during mechanical ventilation, arterial blood pressure, and pulse rate. The patient could also call the nurse for an unplanned visit. The general practitioner visited the patient twice a week. Hospitalization was possible in the case of worsening symptoms or at the patient's request.

## **Statistical Analysis**

The data are reported as mean  $\pm$  SD. We used one-way analysis of variance for breakdown series, and we assumed an absence of difference between variance (Levene test). The differences between 2 mean values were analyzed with Scheffé's test for post-hoc analysis. A p value of < 0.05 was considered significant. Calculations were performed with statistics software (Statistica, Statsoft, Tulsa, Oklahoma).

## **Results**

During the period 1998–2002, 195 of our patients used long-term mechanical ventilation at home. Of those 195 patients, 15 suffered from chronic respiratory failure related to severe idiopathic kyphoscoliosis. Among those 15 patients, 8 had an infection-related ARF and were therefore eligible for this study. Tables 1 and 2 describe these patients' baseline conditions. There were 5 men and 3 women, age  $61 \pm 10$  years, with a Cobb angle of  $84 \pm 7^{\circ}$ ,

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Table 1. Patient Characteristics and Ventilator Settings

Patient	Sex (M/F)	Age (y)	Cobb angle (degrees)	Duration of Mechanical Ventilation (months)	Ventilation Mode	IPAP (cm H <sub>2</sub> O)	V <sub>T</sub> (mL)	Respiratory Rate* (breaths/min)
1	M	45	83	19	NPPV P	20	NA	16
2	F	59	85	30	NPPV V	NA	450	18
3†	F	68	84	18	NPPV V	NA	650	16
4	M	56	78	24	NPPV V	NA	650	16
5	M	62	88	14	NPPV P	16	NA	16
6	M	53	90	108	Tracheostomy V	NA	550	16
7†	F	78	95	23	NPPV P	18	NA	16
8‡	M	67	72	11	NPPV P	16	NA	16
Mean $\pm$ SD	NA	$61 \pm 10$	$84 \pm 7$	$31 \pm 32$	NA	NA	NA	$16\pm0.7$

<sup>\*</sup>Backup rate on pressure-assisted mode and minimum frequency on volume-assisted mode

IPAP = inspiratory positive airway pressure on pressure-cycled mode

 $V_T$  = tidal volume on volume-cycled mode

NPPV = noninvasive positive-pressure ventilation

P = pressure-assisted ventilation

NA = not applicable

V = volume-assisted ventilation

Table 2. Baseline Steady-State Pulmonary Function Test and Blood Gas Values at the Last Regular Hospital Checkup Before This Episode of Acute Respiratory Failure\*

Patient	Months Since Last Hospital Checkup	Percent of Predicted FVC	FEV <sub>1</sub> /FVC (%)	Percent of Predicted TLC	рН	P <sub>aO2</sub> (mm Hg)	P <sub>aCO2</sub> (mm Hg)	HCO <sub>3</sub> <sup>-</sup> (mmol/L)	Mean Nocturnal S <sub>pO2</sub> † (%)	$\begin{array}{l} \text{Nocturnal} \\ S_{pO_2}\text{Time} \\ < 90\% \dagger \\ (\%) \end{array}$
1	4	27	83	40	7.47	84	37	23	95	0
2	6	38	99	50	7.42	69	49	26	91	4.2
3‡	8	46	115	55	7.43	72	45	25	93	3.5
4	4	43	92	52	7.44	85	43	25	96	0
5	5	32	85	47	7.41	73	47	27	94	1.2
6	10	30	105	45	7.43	82	43	24	96	0
7§	6	27	107	56	7.44	69	52	30	92	3.4
8	5	34	102	57	7.41	65	50	28	91	9
Mean ± SD	$6 \pm 2$	$34 \pm 7$	$98 \pm 11$	$50 \pm 6$	$7.43 \pm 0.02$	$75 \pm 8$	$46 \pm 5$	$26 \pm 2$	$93.5 \pm 2$	$4.3 \pm 2.9$

<sup>\*</sup>While spontaneously breathing room air unless otherwise noted

who had been on mechanical ventilation for  $31 \pm 32$  months. All but one were treated with NPPV, with a well-fitted commercial nasal mask (four with a Profile Gel, Respironics, Murrysville, Pennsylvania, and three with a Sullivan, ResCare, San Diego, California). Three used a volume-assist ventilator (Home 1, Airox, Pau, France), and four used a pressure-assist ventilator (Onyx, Pierre Medical, Verriere le Buisson, France), with a circuit

that includes an expiratory valve, and the ventilator was set with a backup rate of  $16 \pm 0.7$  breaths/min. Only one patient was invasively ventilated (LP10, Puritan Bennet, Pleasanton, California). That patient lived at home in stable clinical condition, with no need for hospitalization during 6 months of followup. Table 3 shows the blood gas and oximetry values at the first home visit for and the first night of this ARF episode.

<sup>†</sup>Received supplemental oxygen during mechanical ventilation

<sup>‡</sup>Treated in the hospital

<sup>†</sup>During nocturnal mechanical ventilation during annual hospital checkup

<sup>‡</sup>While receiving supplemental oxygen at 1.5 L/min during spontaneous breathing, and at 2.0 L/min during mechanical ventilation

<sup>§</sup>While receiving supplemental oxygen at 2.0 L/min during spontaneous breathing and mechanical ventilation

<sup>||</sup>Treated in the hospital

FVC = forced vital capacity

 $FEV_1$  = forced expiratory volume in the first second

 $S_{pO_2} = blood$  oxygen saturation measured via pulse oximetry

 $S_{pO_2}$  time < 90% = percent of total sleep time with  $S_{pO_2}$  < 90%, during nocturnal mechanical ventilation

Table 3. Blood Gas and Oximetry Values at the First Home Visit for and the First Night of This Episode of Acute Respiratory Failure

Patient	During Spontaneous Breathing on Room Air, at the First Home Visit for the ARF Episode			Mean S <sub>pO2</sub> During	After 1 h of MV			During Nocturnal MV on the First Night	
	рН	P <sub>aCO2</sub> (mm Hg)	P <sub>aO2</sub> (mm Hg)	1st hour of MV (%)	рН	P <sub>aCO2</sub> (mm Hg)	P <sub>aO2</sub> (mm Hg)	Mean S <sub>pO2</sub> (%)	S <sub>pO2</sub> Time < 90% (%)
1	7.31	69	40	93	7.38	51	71	92	14
2*	7.30	65	51	88	7.39	53	74	91	18
3†	7.29	68	52	90	7.38	55	65	93	9
4	7.32	59	63	95	7.39	48	81	94	7
5*	7.28	67	48	89	7.37	55	67	93	16
6	7.30	64	51	91	7.38	51	73	93	7
7†	7.26	71	40	87	7.37	54	85	91	13
8‡	7.29	70	43	89	7.39	54	71	88	36
Mean ± SD	$7.29 \pm 0.02$	$67 \pm 4$	$49 \pm 8$	$90 \pm 3$	$7.38 \pm 0.01$	$53 \pm 3$	$73 \pm 7$	$92 \pm 2$	$15 \pm 12$

<sup>\*</sup>Began supplemental oxygen

One patient (number 8) refused to be treated at home and he also had a mean nocturnal  $S_{pO_2}$  of 90%, so he was excluded from the study and treated in the hospital. Thus, 7 patients were included, and all of these were successfully treated at home. With patients 3 and 7 we slightly increased the oxygen flow. With patients 2 and 5, who had not been using supplemental oxygen before this ARF episode, we administered oxygen for a few days.

The initial sputum cultures found *Pseudomonas aeruginosa* in patients 2, 3, and 7, and *Streptococcus pneumoniae* in patients 1 and 6. The sputum cultures were negative in the other patients.

Figures 1, 2, and 3 show the ABG results. The pH,  $P_{aO_2}$ , and  $P_{aCO_2}$  values (during spontaneous breathing, after 1 hour off the ventilator) all returned to baseline 4 weeks after the beginning of the ARF.

The patients progressively decreased the daily duration of mechanical ventilation, according to their ability to breathe comfortably without mechanical assistance. After 4 weeks they had regained a steady state, with the same weekly duration of ventilatory assistance as before the ARF episode (Fig. 4).

During the study, neither the nurse nor the general practitioner were called for an emergency visit.

#### Discussion

All seven of these kyphoscoliotic patients on long-term mechanical ventilation, suffering from infection-related ARF, were successfully treated at home.

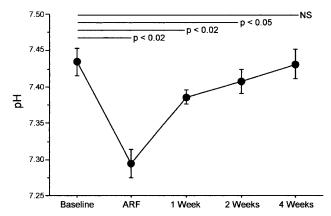


Fig. 1. Mean  $\pm$  SD pH during spontaneous breathing before, during, and after an episode of acute respiratory failure (ARF) in 7 patients treated at home.

Bach et al<sup>13</sup> found that, in neuromuscular patients on home long-term mechanical ventilation, who presented with acute ventilatory failure, increasing the daily duration of mechanical ventilation to 24 h/d, combined with secretion clearance maneuvers, dramatically reduced the need for hospitalization.

In the present study we based our assessment of respiratory tract infection on cough, excessive sputum production, and fever, and the respiratory tract infections caused the ARF in these patients. Obviously, antibiotics were the cornerstone of treatment, and the role of the continuous mechanical ventilation was to support ventilation until the

<sup>†</sup>Increased supplemental oxygen flow

<sup>‡</sup>Treated in the hospital

ARF = acute respiratory failure

SpO2 = blood oxygen saturation measured via pulse oximetry

MV = mechanical ventilation

 $S_{pO_2}$  time <90% = percent of total sleep time with  $S_{pO_2}<90\%$ 

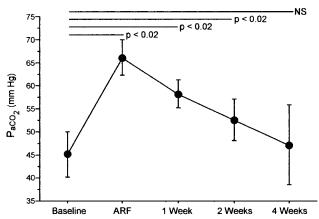


Fig. 2. Mean  $\pm$  SD P<sub>aCO2</sub> during spontaneous breathing before, during, and after an episode of acute respiratory failure (ARF) in 7 patients treated at home.

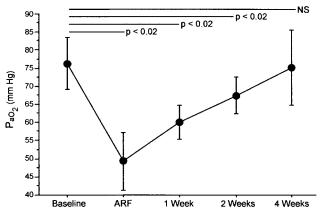


Fig. 3. Mean  $\pm$  SD P<sub>aO2</sub> during spontaneous breathing before, during, and after an episode of acute respiratory failure (ARF) in 7 patients treated at home.

antibiotics took effect. All the patients but one, who was tracheostomized, were treated with NPPV.

NPPV decreases the need for endotracheal intubation and decreases the risk of complications associated with invasive ventilation in patients with acute-on-chronic ventilatory failure. 14-18 Patients in the latter prospective randomized controlled trials exclusively or predominantly had COPD. Patients with thoracic restrictive disorders were also successfully treated with NPPV in open studies 19,20 and in a randomized controlled trial.21 Robino et al22 reported that NPPV was less successful in thoracic restrictive disorders (35%) than in COPD (67%), but 70% of those patients were not on home long-term mechanical ventilation, so the first step was to start NPPV as soon as possible. Starting NPPV takes time because it can be difficult to find comfortable ventilator settings and the correct interface. Starting NPPV during a critical condition is more difficult and takes more time, so there is a higher risk of rapid worsening and the need for intubation.<sup>23</sup> In our

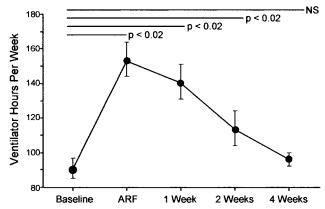


Fig. 4. Mean  $\pm$  SD ventilator hours per week before, during, and after an episode of acute respiratory failure (ARF) in 7 patients treated at home. Before this ARF these patients had been using mechanical ventilation only at night.

study the patients were already established users of long-term home NPPV, so we did not have those difficulties. The positive effect of the patient being accustomed to NPPV was also evident in the management of COPD exacerbation in patients with and without home long-term mechanical ventilation.<sup>24</sup>

In treating acute ventilatory failure it is crucial to provide adequate alveolar ventilation, particularly early in the ARF episode. In the present study this was easy to achieve by asking the patient to continuously use the ventilator, without any change to the ventilator settings. This increase in daily duration of mechanical ventilation was well tolerated. In our protocol we did not change the ventilation mode or settings, which had been previously titrated and regularly re-evaluated for adequate ventilation and the patient's comfort. Our patients also kept using the nasal mask they had been using before the ARF episode, because nasal masks had been successfully used<sup>15–18</sup> and are better accepted than other interfaces,25 though face masks are usually recommended for acute ventilatory failure. Familiarity with the previously used masks and ventilator settings could improve patient adherence to the request that they continuously use their NPPV, which we think partly explains the effectiveness of our home-treatment protocol.

In our study the ARF severity assessment was based on worsening  $P_{aO_2}$ ,  $P_{aCO_2}$ , and pH (acute respiratory acidosis, mean pH 7.29  $\pm$  0.02, compared to baseline pH of 7.43  $\pm$  0.02). All the patients had a rapid positive response: pH,  $P_{aCO_2}$ , and  $P_{aO_2}$  improved significantly (p < 0.02) after 1 hour of mechanical ventilation. This is in agreement with studies that found that rapid (within 2 h) improvement of blood pH was crucial for NPPV success. 17,26–29 The overnight  $S_{pO_2}$  while on mechanical ventilation during the first night was an important element in our evaluation of ARF severity and the decision to treat the ARF at home. Patient 8, who decided to be treated in

the hospital, had the worst overnight  $S_{pO_2}$  while on mechanical ventilation during the first night.

The pH,  $P_{aCO_2}$ , and  $P_{aO_2}$  returned to near baseline in 4 weeks, with the same daily duration of ventilatory assistance as before the ARF episode. Our results are comparable to those from COPD patients in acute ventilatory failure treated in a general ward, <sup>18</sup> even though the time to return to baseline was longer in the present study. This may be related to the fact that our patients had kyphoscoliosis, not COPD. We had the advantage that our patients were already equipped for and accustomed to home mechanical ventilation, so it was relatively easy to increase the daily duration of mechanical ventilation and add or increase the supplemental oxygen in the home setting.

A limitation of this study is the lack of a control group; we cannot state that the same outcome could not have been achieved without any intervention, or with antibiotics only. Nevertheless, our assessment of ARF severity was based partly on the ABG values, which showed respiratory acidosis, and the relatively long time it took to return to baseline. In addition, the conventional hospital treatment for these patients includes antibiotics and increased mechanical ventilatory support, so it would not have been ethically possible to omit those treatments at home.

There is an obvious financial advantage to home treatment of ARF with these patients. We estimate that the average cost per patient of our approach was 1,300 Euros, which includes the costs of the clinician visits, the tests and monitoring (ABG, sputum culture, electrocardiogram, and overnight oximetry), and the drugs and oxygen. The average duration of hospitalization of these patients is 2–3 weeks, which costs 6,500–8,000 Euros per patient, so the home treatment was substantially cheaper.

# Conclusion

With kyphoscoliotic patients on long-term mechanical ventilation, suffering from infection-related ARF, home treatment is feasible, efficient, and cost-effective, provided there is adequate collaboration by the patient and family, and staff well-trained in mechanical ventilation and other aspects of the home care of these patients.

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Nurse visiting patient's home Undated photograph Courtesy National Libraries of Medicine