Non-invasive Mechanical Ventilation Enhances Patient Autonomy in Decision-Making Regarding Chronic Ventilation

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ABSTRACT

Objective: *Patients with respiratory failure due to progressive muscle weakness often require chronic ventilatory support, but many do not make decisions regarding ventilation prior to a crisis. We studied the use of non-invasive ventilation as a tool to enable communication and facilitate decision-making regarding chronic ventilation.*

Methods: *Patients with profound muscle weakness and acute respiratory failure, were supported or weaned by non-invasive positive or negative pressure ventilation. The patients were then interviewed and their informed autonomous decisions were used to plan their future management.*

Results: *Non-invasive ventilation could be used safely to support patients with acute respiratory failure until decisions regarding chronic ventilation are made and as an alternative means of ventilation for those who refuse tracheostomy.*

Conclusions: *Non-invasive ventilation may be used in patients with profound muscle weakness, as a means of enhancing patient autonomy by improving communication and maintaining ventilation until decisions about ongoing care are made.* (**Critical Care and Resuscitation 2005; 7: 116-118**)

Key words: Patient autonomy, non-invasive ventilation, progressive muscle weakness

Respiratory failure requiring long term ventilation may occur in patients with chronic disease such as end stage lung disease[1] or with progressive muscle weakness, such as amyotrophic lateral sclerosis (ALS), where respiratory insufficiency occurs in almost all patients before death.[2-4] In terminal diseases in which respiratory failure is imminent, decisions regarding long-term ventilation should be made in advance, as part of patient autonomy.[1,5] Unfortunately, less than half of patients with "terminal" disease with respiratory failure due to progressive muscle weakness make decisions regarding their management prior to the development of crisis situations.[5]

Respiratory failure may develop in patients with muscle weakness because of abdominal muscle weakness with an inadequate cough, upper airway or bulbar dysfunction with aspiration of secretions, or mucus plugs obstructing airways. When this occurs, the options available to the patient are invasive, chronic mechanical ventilation, non-invasive ventilation or no ventilation at all.[2]

Once patients are intubated in an emergency situation, they may find themselves unable to express their preferences regarding future management, as tracheal intubation and mechanical ventilation render them unable to speak. In awake patients who can write, this may not be such a problem as they may be able to communicate adequately. However, in patients who have profound limb weakness, this may not be possible. Communication with them is then limited to 'yes' and 'no' answers and painstaking spelling of words letter by letter, which may be time-consuming and frustrating.

Patients with respiratory failure due to muscle weakness may also be supported by non-invasive ventilation

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using either positive pressure ventilation (NIPPV) or negative pressure ventilation (NINPV) using Biphasic Cuirass ventilation (BCV). Both modalities have been shown to maintain oxygenation and tidal ventilation in these patients.[2,6,7] Biphasic Cuirass ventilation may also increase expiratory flow, enabling patients to speak and cough in order to clear secretions.[6]

We studied the use of non-invasive ventilation in patients with respiratory failure due to muscle weakness, as a tool to enable patient communication to facilitate decision-making and foster patient autonomy.

The following cases illustrate the use of non-invasive ventilation to foster the autonomy of patients in the acute situation, as well as in more chronic problems requiring long-term ventilation.

Case report 1: A 57 year old man presented with severe polymyositis and had numerous admissions with acute on chronic respiratory failure, requiring intubation and mechanical ventilation (MV). On his first admission his family was advised he would need a tracheostomy because he was classified as having terminal disease with respiratory failure, but he persistently refused tracheostomy when asked. On this and on subsequent admissions he was successfully weaned from MV with NIPPV, by face- and then nose-mask. BCV was tried, but was not suitable for his body habitus (morbid obesity). He was supported by NIPPV every night of his remaining life. He had much fewer admissions for acute on chronic respiratory failure (usually due to intercurrent infection) over the ensuing 4 years. He persistently refused tracheostomy even when towards the end of his life he needed the NIPPV by face-mask for most of the day and night.

Case report 2: A 72 year old male with ALS for 9 years was admitted to the ICU in respiratory failure secondary to severe sepsis from a urinary tract infection. His physical function before admission was limited to bed rest without any ability to move his limbs. He was able to speak until the sudden development of respiratory failure associated with urinary tract sepsis. On admission to the ICU he required intubation and was treated with appropriate antibiotics. On recovery from the sepsis he was fully alert and aware of his condition and was trying to communicate with the attending physicians and nurses. Weaning trials revealed a very poor muscle response, a trial of T-piece weaning resulted in tachypnoea, desaturation and an increase in $PaCO_2$. The patient was put onto BCV negative pressure ventilator, which allowed successful extubation. The patient was then able to communicate verbally, assisted by the expiratory phase of the cuirass ventilation. Neurological and psychiatric evaluation at this time confirmed that the patient had no mental impairment and was fully aware of his condition. The patient repeatedly stated that he wished discontinuation of all modes of mechanical ventilation, and reported that the suffering and hopelessness of his disease was worse than his previous experience in a concentration camp. Therefore, cuirass ventilation was discontinued at the patient's insistence and he was given small analgesic doses of morphine for pain relief. He died comfortably 36 hours after ventilation was discontinued

Case report 3: A 62 year old man with ALS was admitted to our ICU with respiratory failure due to complete atelectasis of the left lung. He was intubated and ventilated enabling bronchoscopy to remove mucus plugs. He underwent subsequent extubation and was supported by BCV, but unfortunately developed repeat atelectasis and needed re-intubation. On questioning prior to re-intubation he consented to undergo a tracheostomy and has been ventilated since for about 5 years. He has since written 4 books - including one on aspects of euthanasia.

Case report 4: A 78 year old female with progressive muscle weakness required repeated admissions for mechanical ventilation. On admission to our unit she presented with atelectasis and was intubated and ventilated. Two extubation attempts failed until she was put on BCV and weaned successfully. When asked, she refused tracheostomy and was later sent home where she continued to be ventilated non-invasively by BCV for several more years.

Case report 5: A 30 year old man presented with sudden quadriplegia. Clinical imaging revealed a large cervical epidural abscess. Upon further questioning it was found that he was an intravenous drug user and the abscess was thought to be secondary to bacteremia due to intravenous drug use. Drainage of the abscess did not result in neurological improvement. The spectre of lifelong mechanical ventilation was then realised. As the patient was unable to move his limbs or indeed nod or shake his head it was not possible to ascertain his views regarding his prognosis and management. He was therefore transferred from intermittent positive pressure ventilation via tracheostomy tube to biphasic cuirass ventilation. With occlusion of the tracheotomy tube during exhalation, with the cuff deflated, he was able to vocalise. This enabled discussion of ongoing management with the patient's consent and involvement.

DISCUSSION

In order to honour patient autonomy in patients who suffer from incurable diseases, physicians should discuss treatment options and preferences while patients are still competent and able to communicate.[8] However, it has been shown that patient-physician discussions and definitive decisions are lacking, especially in these

situations.

Patients who are expected to progress to end stage respiratory failure should be informed and encouraged to make decisions regarding ventilation prior to a crisis.[3,5] Unfortunately, as many as 68% of ALS patients do not make decisions regarding ventilation prior to the inevitable respiratory failure.[3]

In those cases that have not made advanced directives prior to the crisis, the issue of autonomy regarding future options may be complicated by a reduced ability to communicate because of an endotracheal tube, decreased bulbar function, and/or severe muscle weakness preventing them from writing. Also, relying on a family member or surrogate to make decisions in this setting is unwarranted, especially as most of these patients are fully awake.[8] It is therefore important to try and improve patients' communication ability using non-invasive techniques.

Non-invasive ventilation has been used in cases of respiratory failure due to progressive muscle weakness, provided upper airway protective mechanisms are maintained and lung compliance is reasonable.[2,6,9] NIPPV has been shown to increase patient survival, with favourable results on respiratory function and quality of life in those patients with ALS who tolerate it.[9]

External biphasic cuirass ventilation can serve as a non invasive method of ventilation in ALS patients, with adequate bulbar function.[6] In awake, co-operative patients, it may improve quality of life, as it allows them to talk and eat. BCV may also be used to help clear secretions, by using cycles of high frequency oscillations followed by several positive expiratory pressure breaths.[6] BCV can also be used as a weaning technique - as biphasic extrathoracic pressures are applied, pressure support levels required for adequate ventilation may be reduced until extubation is feasible.[6,10]

As we have shown in the case series above, non-invasive ventilation can be used in patients with respiratory failure due to muscle weakness as a tool to enable earlier extubation or postpone the need for tracheostomy, thus enabling communication with the patients and assisting informed autonomous decisions. It may also provide an alternative, although usually a temporary treatment modality, for those patients who refuse invasive ventilation.

Non-invasive ventilation can also be used to ventilate terminal patients at home, as well as in chronic facilities, thus enabling the patient, family and the treating physician to reconsider all treatment options in a timely manner (an option not always available when intubation and mechanical ventilation are required acutely).

We conclude that non-invasive ventilation using BCV could be tried in ALS patients and other patients with muscle weakness, as a means of enhancing patient autonomy by improving communication and maintaining ventilation until decisions about ongoing care are made.

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