Long term nasal ventilation

First proposed some 10 years ago as an aid for patients with Duchenne muscular dystrophy, positive pressure ventilation administered via the nasal route has become the preferred method of delivering ventilatory assistance to patients with many varieties of chronic respiratory failure. The rapid rise in popularity of nasal ventilation has been fuelled by the advantages of convenience, comfort, and portability over other modes of non-invasive ventilation. In comparison with invasive positive pressure ventilation, non-invasive ventilation eliminates the need for tracheostomy care and suctioning, greatly facilitating the delivery of home mechanical ventilation and substantially reducing costs.¹

Many uncontrolled studies have shown that nasal ventilation reduces symptoms of hypoventilation and improves daytime gas exchange in patients with restrictive thoracic disorders such as slowly progressive neuromuscular syndromes and kyphoscoliosis.²⁻⁴ In addition, nasal ventilation has been shown to ameliorate the obstructive sleep apnoeas and nocturnal desaturations associated with use of negative pressure ventilation,⁵ and to ameliorate the nocturnal hypoventilation and sustained oxygen desaturations that occur commonly in patients with severe neuromuscular and chest wall disorders.⁶⁷ Despite these favourable results, however, evidence to support the widespread, long term use of nasal ventilation has been limited by a lack of long term follow up studies.

With the publication of the study by Simonds and Elliott on pp 604–609 of this issue of Thorax,⁸ and a similar study from France by Leger and colleagues⁹ within the past year, part of the information gap is being closed. The study by Simonds and Elliott provides data on 180 patients with various causes of respiratory failure treated with nasal ventilation at the Royal Brompton Hospital between 1987 and 1992. The French study followed 276 similar patients also for a five year period. Both studies used continuation of nasal ventilation rather than survival as the major outcome variable, but most patients who failed to continue nasal ventilation died. The findings were remarkably similar; patients with restrictive thoracic disorders had highly favourable continuation (and hence survival) rates, ranging from 80% to 100% over the follow up periods. The only exception was the subgroup of patients with Duchenne muscular dystrophy in the French study who had a continuation rate of 56%. In contrast to the favourable findings among patients with restrictive defects, patients with chronic obstructive disorders fared much less well in both studies, with continuation rates of 40-50%. Among patients with bronchiectasis who were often on lung transplantation lists, most failed to survive the initial two years.

In addition to the actuarial data, Simonds and Elliott provide follow up arterial blood gas data which show improved oxygenation and alveolar ventilation in patients with both restrictive and obstructive disorders, although the improvement was slightly smaller in the latter. In a small subgroup of patients switched from negative pressure to nasal ventilation they observed improvements in blood gas tensions following the switch. In addition, results of a quality of life survey are presented which showed that patients receiving nasal ventilation compared favourably with UK population norms and patients in the USA with chronic disorders with regard to mental health, energy, and vitality.

Before firm conclusions are drawn from the study by Simonds and Elliott a number of limitations should be considered, most of which were acknowledged by the authors. The major limitation is the lack of a control group. The authors argue that a randomised prospective study with an untreated control group would be unethical. This argument is valid for most forms of restrictive thoracic disease because the survival results are so favourable, and studies show deterioration in nocturnal gas exchange and symptoms when such patients are temporarily withdrawn from their ventilators.⁶⁷ However, studies comparing nasal ventilation with invasive positive pressure ventilation or even other forms of non-invasive ventilation might not be considered unethical. In addition, subgroups of patients with less favourable results on nasal ventilation, such as those with obstructive lung disease, could still be studied ethically in prospective randomised trials.

Another limitation is the lack of any data on nocturnal gas exchange or sleep quality. The authors surveyed sleep quality and monitored hours of use of the ventilator, but no conclusions about the sustained "correction" of nocturnal hypoventilation are warranted. In addition, the analysis of gas exchange in patients switched from negative pressure to nasal ventilation was inadequate without consideration of hours of daily use and optimisation of pressures. Nasal ventilation has a number of advantages over negative pressure ventilation, but several studies during the 1980s showed that negative pressure ventilation is quite effective at improving gas exchange in patients with chronic respiratory failure due to restrictive thoracic disorders. Without a properly designed trial one cannot conclude from the current data that nasal ventilation is necessarily more effective in improving gas exchange. Additionally, the authors acknowledge that the results of the quality of life survey are difficult to interpret without a baseline analysis or control group. The authors conclude that patients using nasal ventilation have a better quality of life than some investigators have previously thought. However, other investigators have found that over half of chronically ventilated patients with Duchenne muscular dystrophy are very or somewhat satisfied with their lifestyle.¹⁰ In this latter study family members registered more dissatisfaction with lifestyle limitations than the patients themselves, and in future studies quality of life should be assessed not only in patients and appropriate control groups, but also among family members and caregivers.

Despite the above limitations, the conclusion of Simonds and Elliott that results of nasal ventilation among patients with restrictive thoracic disorders are "encouraging" seems well justified. This finding is compatible with that of the French study⁹ and also an earlier French study reporting long term follow up results on patients using invasive positive pressure ventilation.¹¹ In this earlier study, postpolio, kyphoscoliotic, post-tuberculosis, and myopathic patients had five year survivals of approximately 95%, 75%, 70%, and 65%, respectively, suggesting that survival rates among similar patients using nasal ventilation compared favourably with or exceeded those obtained using invasive positive pressure ventilation. However, a direct trial that compares these two modalities in patients with restrictive thoracic disease is unlikely to be performed because, justifiably, few patients would be willing to con596

sider invasive positive pressure ventilation if they are good candidates for nasal ventilation.

Lest we interpret the findings of Simonds and Elliott as an endorsement of the widespread use of nasal ventilation among all patients with chronic respiratory failure, a number of caveats must first be borne in mind. Firstly, Simonds and Elliott selected only patients with intact bulbar function. Patients with chronic respiratory failure due to neuromuscular syndromes associated with bulbar dysfunction would undoubtedly have much poorer survival rates and, if such patients desire prolongation of survival, invasive positive pressure ventilation is the preferred ventilatory mode. In addition, the specified aetiology for chronic respiratory disease must be considered with regard to long term efficacy. Clearly, patients with chest wall disorders and with very slowly progressive neuromuscular syndromes such as post-polio syndrome remain stable on nasal ventilation for long periods of time and are appropriate candidates. This is not the case, however, for patients who have obstructive lung disorders, as Simonds and Elliott point out. It cannot be concluded from their data that these patients fared better or lived longer because of nasal ventilation, despite the improvement in gas exchange during use. Other recent studies suggest that patients with chronic obstructive pulmonary disease (COPD) with substantial daytime hypoventilation and nocturnal breathing disturbances may benefit from nasal ventilation,¹² but prospective, randomised long term trials will be necessary to show convincingly that nasal ventilation with oxygen supplementation achieves sustained improvements in daytime gas exchange, quality of life, and survival in these patients compared with oxygen supplementation alone.

Another subgroup of patients that deserves closer scrutiny is that of the more rapidly progressive neuromuscular syndromes such as Duchenne muscular dystrophy. Simonds and Elliott did not analyse them separately, but the French study⁹ found that these patients continued nasal ventilation less often than other patients with neuromuscular disease, more often necessitating tracheostomy. In addition, Raphael et al¹³ recently reported increased mortality due to respiratory failure among patients with Duchenne muscular dystrophy randomised to use nasal ventilation prophylactically. Although this study had a number of important methodological problems, including more patients with left ventricular dysfunction in the control group and no evaluation of patient compliance, the results nonetheless raise questions about the safety of nasal ventilation among patients with Duchenne muscular dystrophy. The concern raised was that patients receiving nasal ventilation gained a false sense of security and delayed seeking medical assistance during respiratory infections until crises occurred. In view of this concern studies examining the more systematic use of techniques to assist the removal of airway secretions or the earlier institution of invasive positive pressure ventilation among patients with Duchenne muscular dystrophy who value prolongation of survival may be indicated.

Despite these caveats, the study by Simonds and Elliott provides valuable information on how patients with particular types of chronic respiratory failure fare with long term nasal ventilation. The study clearly shows that patients with chest wall deformities and slowly progressive neuromuscular diseases are safely and effectively supported for years with nasal ventilation. However, many questions remain unanswered. As discussed above, more studies are necessary to evaluate the safety and efficacy of nasal ventilation in particular subgroups, such as those with COPD or Duchenne muscular dystrophy. Serial data are lacking on whether the amelioration of nocturnal hypoventilation by nasal ventilation is sustained, or whether deterioration occurs necessitating further adjustments. Some investigators have suggested that alternative interfaces to nasal ventilation such as mouth or oronasal masks may sometimes be more efficacious,¹⁴ and comparative studies would be helpful. In addition, although results were quite favourable with mainly volume ventilation in the study by Simonds and Elliott, some investigators favour pressure support ventilation and direct comparisons of these modes are desirable. Ultimately, the goal of non-invasive positive pressure ventilation should be not only to augment alveolar ventilation, but also to optimise functional status, quality of life and survival, and future studies must also pay close attention to these outcomes.

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