Home ventilation

The report by Brooks et al (pages 99 to 106) in this issue of the *Canadian Respiratory Journal* brought back an instructive memory. Many years ago, I was asked to review a paper for *The American Review of Respiratory Disease* concerning home ventilation (1). The paper was from Saskatoon, and the first author was Vern Hoeppner, who is still there. Obviously, it was accepted for publication. The paper concerned home nocturnal ventilation of patients with kyphoscoliosis. When I picked the paper up, I was incredulous. The paper amounted to a (small) series of case reports indicating that home ventilation worked. There were no controls. The outcomes were essentially measurements of blood gases and what we now call 'quality of life', including hospitalization records. At first glance, it looked like a report by enthusiasts who were recounting anecdotes. I then read the paper – and I believed. Hoeppner and colleagues reported on a group of patients with end-stage respiratory failure due to kyphoscoliosis, in whom essentially nothing worked, including home oxygen (then a relatively new therapy). These people underwent a remarkable improvement, especially in daytime blood gases, with nocturnal ventilation. They went from being more or less permanent hospital residents to living independently at home. I had had enough clinical experience with this type of patient to recognize that this was an important result that was most unlikely to be explicable on the basis of anything but home ventilation. At that time, in my view, there was no need for a randomized clinical trial of home ventilation; this was sort of like penicillin in pneumococcal pneumonia.

The paper by Brooks et al strongly supports my clinical judgement and that of Vern Hoeppner, who had the idea first. In an apparently carefully selected group of patients with respiratory failure due to kyphoscoliosis and neuromuscular disease, they show, over years of study, remarkable stability and survival during treatment with home ventilation, which was largely nocturnal. Improvements in sleep quality, blood gases and 6 min walking distance were all extremely well maintained. Some of the patients with kyphoscoliosis had been followed up for 10 years, with little discernable deterioration. This is striking in a group with an average forced vital capacity of 27% of the predicted normal value at the time of study entry. The patients with neuromuscular disease have not been followed up for as long, and will almost certainly not do as well, because in many of these patients, the diagnoses

imply progressive disease. In any event, the paper by Brooks et al strongly supports the use of home ventilation in people with respiratory failure due to thoracic restrictive or neuromuscular diseases. In Manitoba, there is a provincially funded program to supply this therapy. This and other programs should be able to produce results similar to those of Brooks et al, if they are given adequate levels of training and home support.

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There are, I note, still mysteries associated with the success of home ventilation. The first mystery is the ability of nocturnal ventilation to produce long term improvements in daytime blood gases, which I believe is critical to the success of the therapy. The deep breaths necessitated by nocturnal ventilation may produce improvements in the mechanics of the respiratory system, permitting more ventilation per unit of neural drive. This has been well shown in short term experiments, but I would be surprised if such improvements persisted for 12 h. It is possible that nocturnal ventilation rests the muscles of inspiration, which would otherwise malfunction because of chronic fatigue. If this is the case, it is the only instance of chronic fatigue of these muscles of which I am aware. Finally, it is possible that chronic carbon dioxide retention reduces ventilatory drive in these people, and that nocturnal ventilation, by lowering the carbon dioxide level, allows the respiratory centre to reset, so that the drive during the day is closer to normal. To me, this explanation is the most attractive, but I do not know of firm evidence that supports this explanation over the others. In terms of the patients, the explanation does not matter. A mystery introduced by the paper of Brooks et al is the failure of lung function to decline much with time in their patients with kyphoscoliosis; over 10 years, the vital capacity declined by 5.8%! This is much less than one would expect on the basis of normal aging. In another series, it would be easy to explain this as a survivor effect; that is, that lung function could not decline with survival. Therefore, of course, those who sur-

vived would have little to no decline. However, this explanation assumes a substantial nonsurvival rate, and only two of the patients with kyphoscoliosis in the study by Brooks et al died. They should continue to follow these patients to see if the remarkable stability of lung function continues as more patients reach the five- to 10-year follow-up mark.

Finally, I believe that this story is instructive in that it indicates again that, in some circumstances, randomized, controlled clinical trials are superfluous. When the outcome of a disease is well known, and a particular therapy changes the outcome drastically, further examination is not needed. Now, we only need to be able to define 'well known' and

'drastically'. Both are, I fear, subject to interpretation, but can be applied to respiratory failure in kyphoscoliosis and home ventilation, as Vern Hoeppner recognized so long ago.

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REFERENCES

1. Hoeppner VH, Cockcroft DW, Dosman JA, Cotton DJ. Nighttime ventilation improves respiratory failure in secondary kyphoscoliosis. *Am Rev Respir Dis* 1984;129:240-3.