You Need More Than Nocturnal NIPPV to Manage Duchenne’s Muscular Dystrophy

To the Editor:

Leger et al reported that the use of predominantly nocturnal nasal intermittent positive pressure ventilation (NIPPV), 10.5 ± 2 h/d, can improve arterial blood gas values, reverse symptoms of alveolar hypoventilation, and decrease hospitalization rates for patients with intrinsic lung disease, kyphoscoliosis, and Duchenne’s muscular dystrophy (DMD). Only 35% of their DMD patients continued NIPPV for 3 years. The others either voluntarily stopped treatment, died, or required tracheostomy IPPV. Except for two patients with aerophagia, the authors did not indicate why patients voluntarily stopped treatment, why they died, or what was meant by them requiring a tracheostomy.

As in our program in earlier years, Leger et al neither provided adequate daily periods of noninvasive IPPV nor effectively cleared airway secretions noninvasively when necessary. In reality, the 10.5 h/d of NIPPV could never be sufficient to sustain DMD patients indefinitely. Intercurrent upper respiratory tract infections with nasal congestion and airway secretions necessitate 24 h noninvasive IPPV, often by means other than NIPPV. Mucus plugging, which the patients cannot clear because of inability to generate cough flows of at least 3 to 5 L/s, needs to be managed by manually and mechanically assisted coughing. It is not surprising, therefore, that most of Leger’s DMD patients were hitherto intubated, tracheostomized, or deceased. Further, by providing “supplemental oxygen … during NIPPV and time off … to maintain the SaO2 >90%,” they permitted the DMD patients to underventilate and to be encumbered with secretions at critical times and were bound to fail.

DMD differs fundamentally from the other Leger diagnostic groups. Providing a part-time, predominantly nocturnal, treatment to attain positive results is inadequate when dealing with rapidly progressive primary ventilatory impairment. Although control studies may be useful for determining how nocturnal NIPPV might temporarily affect the evolution of ventilatory deterioration, daytime blood gases and hospitalization rates, these are secondary considerations for patients who in a few years will retain less than 10% of predicated vital capacity and require 24 h use of noninvasive aids to remain healthy and unencumbered by an indwelling tracheostomy.

We managed 52 24-h/d DMD ventilator users, for a mean of 7 years, and in one case from age 18 to 42 years, by noninvasive ventilatory support. Only one patient ever required tracheostomy. Only when bulbar dysfunction entirely precludes paroaral nutrition should tracheostomy be considered for any neuromuscular ventilator user. I suggest that these authors, and others who equate noninvasive positive pressure ventilation with nocturnal NIPPV, use adequate periods of noninvasive IPPV by mouthpiece, oral-nasal interface, or other custom interfaces and eliminate secretions with cough-like expiratory flows before they conclude that nasal, or worse, noninvasive IPPV has failed.

Also, of the more than 340 noninvasive IPPV users we have kept free of tracheostomy, we have had to discontinue noninvasive IPPV and resort to a body ventilator because of aerophagia for only one patient. Aerophagia is a difficult but insurmountable problem.

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REFERENCES

4 Bach JR, Alba AS, Saporito LR. Intermittent positive pressure ventilation via the mouth as an alternative to tracheostomy for 257 ventilator users. Chest 1993; 103:174-82

To the Editor:

Much of what Dr. Bach has written in his letter misquotes and misrepresents our article. For example, the “35%” quoted in the article refers to the probability for Duchenne’s muscular dystrophy (DMD) patients to continue nasal intermittent positive pressure ventilation (NIPPV); none of these patients voluntarily stopped treatment (Ches 1994; 105:100-05). The reasons for stopping treatment are listed and include the reasons for tracheostomy IPPV, “abdominal bloating and distention during NIPPV (n=3), anxiety because of fear that the system would become dislodged during sleep (n=1), . . . and severe hypercarbia during NIPPV use (n=1),” and causes of death, “died during NIPPV due to a respiratory problem” (Ches 1994; 105:100-05). Finally, as stated in the article, supplemental oxygen was not necessary for any of the DMD patients. In addition, it was not the purpose of this study to include secretion clearance techniques and we did not have problems with secretion retention in this group of DMD patients that we studied.

We recognize that there are potential problems of tracheostomy and that is precisely why our preferred method of ventilation is NIPPV. However, we strongly believe that it is very wrong to give the message that tracheostomy is a bad method for long-term ventilation. There are many patients who do very well with tracheostomy ventilation, including many DMD patients. Not all patients are followed up by teams specialized in noninvasive techniques for assisted ventilation; it is often better to do tracheostomy early and provide good ventilation rather than take the risk of doing nothing and possibly be too late to support ventilation in the home.

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Errata

The letter by Dr. Robert Carlen, entitled “Bayes Theorem: Diagnostic Testing for Tuberculosis in Children” and found in the August 1994 issue of Chest (Ches 1994; 106:655-56), should read: “In analogous fashion, the predictive value of a negative result is calculated a priori and is defined as the proportion of all those with a negative result who are free of disease, i.e., the probability of a true negative result divided by the sum of the probabilities of true negatives and false negatives [TN/(TN+FN)].”

“In The Evaluation of Pleural Effusion” by Bartter et al (Ches, 1994; 106:1209-14) the sentence in column two, page 1210 should read, “They found that a gradient <1.2 mg/dl indicated an exudate and a value >1.2 mg/dl indicated a transudate.”