Home Mechanical Ventilation in Amyotrophic Lateral Sclerosis Patients Is Not Always a Problem

To the Editor:

In an editorial in the New England Journal of Medicine concerning assisted suicide and alternatives for patients with amyotrophic lateral sclerosis (ALS),1 L.P. Rowland reported that “the progressive paralysis leads to increasing loss of function, culminating in complete dependence on the help of others for all activities of daily living and, if life is sustained by assisted ventilation, loss of the ability to communicate or swallow.” This sentence is misleading because it implies that the loss of ability to communicate or swallow is caused by the assisted ventilation rather than by the progressive paralysis that occurs as survival is prolonged.

In ALS, speech and swallowing difficulties are directly related to the disease itself. A large number of ALS patients experience swallowing problems and have severe difficulties talking even before ventilatory support is required. Tracheostomy ventilation can exacerbate these difficulties in some patients (but not all, because insufflation leaks with a deflated cuff, allowing speech). In our experience, noninvasive ventilation (NIV) can facilitate speech and swallowing. It also relieves hyperventilation symptoms and can result in weight gain.

Unfortunately, Rowland does not discuss NIV benefits and fails to discuss noninvasive ventilatory options other than remarking that “positive pressure nasal ventilation is an alternative to tracheostomy for some patients.”3 Does he mean tracheostomy is needed only for secretion management and protection against aspiration, or as an alternative to NIV even for patients who require 24-h ventilatory support? In fact, some patients whose time away from ventilation is negligible prefer NIV support to tracheostomy ventilation for comfort, appearance, facilitation of speech, and swallowing. NIV support includes not only nasal ventilation but mouthpiece ventilation, pneumobelt, and other noninvasive muscle aids, and this may significantly prolong life without resorting to tracheostomy.2 Of course, when NIV fails, physicians need to offer the best choices in conventional ventilatory procedures to patients.2

Unfortunately, most physicians and most patients are misinformed about noninvasive (and invasive) methods of prolonging life. Although these guidelines were fairly comprehensive, they failed to address the prevention of deep venous thrombosis (DVT) and pulmonary embolism (PE) occurring among patients with coagulopathy from end-stage liver disease.

A 57-year-old man was admitted to our Medical ICU (MICU) for refractory ascites. He had been suffering from alcoholic cirrhosis for 2 years, with a baseline serum albumin of 2.8 g/dL, total bilirubin of 1.4 mg/dL, and an international normalized ratio of 1.3. During his MICU stay, he required large-volume paracentesis every 48 to 72 h for massive ascites. Other acute problems included upper GI bleeding from esophageal varices that were treated with banding, and nonspecific colitis, which resolved with nasogastric suctioning and rectal tube drainage. With improvement of these problems, he was subsequently listed for liver transplantation. He had been on intermittent pneumatic compression stockings throughout his MICU stay. On the 23rd hospital day, he developed respiratory distress and severe hypoxemia requiring intubation and mechanical ventilation. A lung perfusion scan showed multiple perfusion defects consistent with PE, so an inferior vena cava filter was placed. However, the patient died 2 days later, after his wife requested the withdrawal of life support.

This case demonstrates that PE may occur among patients who are supposed to be auto-anticoagulated due to their liver disease. It also illustrates that intermittent pneumatic compression may not be adequate for DVT prophylaxis among patients with advanced liver disease. To date, there are no available alternatives to nonpharmacologic prophylaxis for venous thromboembolism in these patients. A search for a safe and effective prophylactic modality for DVT and PE is much needed to improve the outcome of end-stage liver patients awaiting transplantation.

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Pulmonary Embolism in a Patient With Coagulopathy From End-Stage Liver Disease

To the Editor:

The American College of Chest Physicians has published guidelines on the prevention of venous thromboembolism.1 Although these guidelines were fairly comprehensive, they failed to address the prevention of deep venous thrombosis (DVT) and pulmonary embolism (PE) occurring among patients with coagulopathy from end-stage liver disease.

Virtually all patients with end-stage liver disease have some component of coagulopathy due to hepatic synthetic dysfunction with failure to manufacture coagulation factors II, VII, IX, and X.2 Meanwhile, they are also at risk for DVT and PE because of an inability to synthesize anticoagulating factors such as protein C, protein S, and antithrombin III.3 The following case summary illustrates these coagulopathic abnormalities.

A 57-year-old man was admitted to our Medical ICU (MICU) for refractory ascites. He had been suffering from alcoholic cirrhosis for 2 years, with a baseline serum albumin of 2.8 g/dL, total bilirubin of 1.4 mg/dL, and an international normalized ratio of 1.3. During his MICU stay, he required large-volume paracentesis every 48 to 72 h for massive ascites. Other acute problems included upper GI bleeding from esophageal varices that were treated with banding, and nonspecific colitis, which resolved with nasogastric suctioning and rectal tube drainage. With improvement of these problems, he was subsequently listed for liver transplantation. He had been on intermittent pneumatic compression stockings throughout his MICU stay. On the 23rd hospital day, he developed respiratory distress and severe hypoxemia requiring intubation and mechanical ventilation. A lung perfusion scan showed multiple perfusion defects consistent with PE, so an inferior vena cava filter was placed. However, the patient died 2 days later, after his wife requested the withdrawal of life support.

This case demonstrates that PE may occur among patients who are supposed to be auto-anticoagulated due to their liver disease. It also illustrates that intermittent pneumatic compression may not be adequate for DVT prophylaxis among patients with advanced liver disease. To date, there are no available alternatives to nonpharmacologic prophylaxis for venous thromboembolism in these patients. A search for a safe and effective prophylactic modality for DVT and PE is much needed to improve the outcome of end-stage liver patients awaiting transplantation.

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Humidification During Continuous Positive Airway Pressure Therapy

To the Editor:

The study by Massie et al has, for the first time, presented scientific proof that the prophylactic application of a humidifier as an accessory in continuous positive airway pressure (CPAP) therapy can improve not only patient compliance and comfort during treatment, but also the results of the treatment. We consider the study to be of particular importance because it shows that we now have to rethink the current clinical practice of using humidifiers as accessories only when patients undergoing CPAP treatment encounter such problems as, for example, drying out of the upper airways.

In their “Discussion” section, the authors maintain that their study is the first to show the effect of humidification on side effects and compliance in obstructive sleep apnea (OSA) patients using CPAP (page 408, lines 34–36). However, our own working group had already published a prospective study on OSA patients experiencing side effects of CPAP treatment (January 1999), in whom the use of a heated humidifier (incidentally, the same make as in the study of Massie et al) both lowered the rate of side effects and improved patient compliance and comfort under treatment. In addition, we were also able to show that the use of a heated humidifier can reduce the number of patients who abandon CPAP treatment because of the above-mentioned side effects.

The results published by Massie et al are in good agreement with our own data, and taken together, our findings now demonstrate the clinical efficacy of heated humidifiers used as accessories in CPAP treatment in terms of improving compliance and treatment comfort as well as the results of treatment.

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