Beyond Mechanical Ventilation for Neuromuscular Patients

Once a neuromuscular patient agrees to prolong life through mechanical ventilation, it is the rehab professional’s responsibility to ensure the patient is introduced to technology that can improve quality of life.

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Photography by Chuck Potter

The use of ventilators (respirators) to extend the lives of patients with progressive neuromuscular diseases has been increasingly undertaken at various medical centers throughout the United States and Europe. To many, the decision to prolong life with a machine is controversial and issues of quality of life are always brought to mind. This controversy is not easily resolved since there is no right answer. The physician’s opinion in this matter is often of dubious value. What matters most is that the patient and his family be given the appropriate information and support to allow them to decide whether to prolong life with mechanical ventilation. The medical staff caring for these patients should be well informed as to the signs of impending respiratory failure in these patients and the treatment modalities for intervention if the family so desires.

The pathophysiology of respiratory failure in the neuromuscular patient is not caused by parenchymal lung disease. It is because of the failure of the respiratory muscle to sustain adequate ventilation. The downhill stages of Duchenne’s muscular dystrophy are unfortunately fairly easy to predict. The same is true for the respiratory problems these patients have.

The vital capacity has been shown to be useful in differentiating subgroups of patients with Duchenne’s muscular dystrophy according to their respiratory ability. Vital capacity less than 1200 ml is associated with more extremity weakness and earlier death around 17 years. Patients with values between 1200 ml and 1700 ml do better clinically and usually live to 20 years. Another predictive value of the vital capacity has been in forecasting the onset of impending respiratory failure. A patient with a vital capacity of 500 ml usually will go into respiratory failure within six months of achieving this level.

By following the vital capacity, respiratory failure is usually predictable in this disease. Clinically alone, however, it is a subtle entity to predict. The onset of respiratory failure is insidious and often sneaks up on the patient. Patients often complain of restless nights, and have morning headaches. This may be attributed to the discomfort of not being able to move and having to be turned frequently throughout the night. Eventually, breathing and
swallowing may be difficult to perform together and, consequently, eating is abandoned and the patient will lose weight.

Before the patient has reached the point of impending respiratory failure, it is necessary to explore with the family and the patient decisions about extending life with a respirator. In our Neuromuscular Clinic at Rancho Los Amigos Medical Center, we have established a special clinic to address the needs of the older patient with Duchenne’s dystrophy.

The goals of the clinic are twofold. The first is education. In our clinic both the family and the patient are educated in every aspect of the respiratory disease. Questions are answered, and the opinion and decision of allowing for a natural death or prolonging the life of the patient are respected. The second goal, then, is to help the patient obtain either transition as smoothly as possible. In the patient desiring natural death that would entail psychological support as well as prevention of any unwanted intervention. For the patient desiring respirator assistance, the goal would be to offer that assistance electively before the onset of acute respiratory failure, which would necessitate an emergency intubation.

Different facilities have chosen different forms of ventilator support. The use of negative pressure systems, such as tank respirators or body cuirases, obviously can be applied without the use of a tracheostomy. On the other hand, they are not portable and cause significant restrictions on functional activities. They are useful initially for nighttime ventilation only. Various modifications in positive pressure masks or the new in-vogue nasal mask or nasal pillows are some other methods used, but again these systems are most useful for nighttime ventilation. Once the patient loses his ability to cough and clear his secretions, tracheal positive pressure ventilation is required. At our facility the transition to respiratory support has been uncomplicated in the majority of these patients. As a whole, they have fewer pneumonias than they had prior to assisted ventilation. They spend very few days sick and usually are up in their powered wheelchairs and able to continue at their usual activities. Most of our patients are discharged home.

Another neuromuscular disease which commonly requires ventilator support is Spinal Muscular Atrophy. In respiratory failure associated with this disease, the diaphragm muscle is relatively spared compared to the intercostal muscle, and patients develop a characteristic “Christmas Tree” deformity of the thorax. Patients with this disease are highly susceptible to pneumonias that might collapse the lungs.

Patients with Spinal Muscular Atrophy are chronic hyperventilators. When healthy and well, they breathe with shallow rapid respirations, just getting by, until a respiratory infection occurs and they rapidly progress to respiratory failure. Unfortunately, the onset of respiratory failure is not easy to predict in these patients and one has to be prepared for acute intervention.

Tracheostomies and ventilator support have added a great deal in prolonging the life of these patients. In our clinic, about 15 patients with the intermediate form of the disease have been placed on tracheal positive ventilation. The use of a tracheostomy in these patients has made it possible to clear secretions and prevent the development of a pneumonia. The respirator, too, has aided these patients. Usually the patient is started on a respirator as an emergency for an acute pneumonia.

After their pneumonia resolves, weaning the patients from full-time ventilator support to only nighttime support was initially possible in all cases. The use of the respirator at night allows the weak respiratory muscle a rest, thereby preventing total fatigue and lung collapse to occur. We have found nighttime ventilation in this patient population to be very successful, and have commonly seen patients not require full-time support for upwards of 10 years after initial ventilation has begun.

Most of our ventilator dependent children live at home. Before the child is sent home, the family is fully trained in all aspects of the child’s care. Since the patients are fully dependent on their caretakers by the time they require ventilator assistance, their quality of life does not change dramatically with the use of the respirator.

Modern technology has opened many new frontiers for the ventilator-dependent neuromuscular patient. Respirators fit easily below electric wheelchairs and are equipped with long-acting batteries. This allows access to the community and the world outside the protective hospital walls. Assistive devices such as mobile arm supports allow for creative expression. The world of computers has allowed these patients a mode of recreation, environmental controls and employment opportunities.

For our patient population, we have found that the majority of the patients and their families choose to prolong their lives with mechanical ventilation. Most make the transition to respiratory support without complications, and it is often demonstrated that their overall clinical condition improves. We have shown that home care of the respirator-dependent patient works, but new opportunities bring with them always new responsibilities. As the number of ventilator-dependent neuromuscular disease patients continues to increase, we will have to meet the challenge of improving the quality of life for these patients.  

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