Home Care Utilizing a Ventilator in a Patient with Amyotrophic Lateral Sclerosis

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A patient with respiratory neuromuscular paralysis secondary to amyotrophic lateral sclerosis is described who was managed at home for more than 12 months on a volume cycled respirator. Although the cost is high, it is less than in an acute care hospital setting. Requirements for success include patient selection, adequate home environment, and dedicated medical and paramedical personnel.

Amyotrophic lateral sclerosis (ALS) is a well known but uncommon neuromuscular disease, predominately affecting middle age males especially in their ability to move, speak, and breathe.[1] Respiratory problems usually occur late in the disease, often secondary to bulbar paralysis. However, there are cases of ALS in which dyspnea or respiratory failure were the initial symptoms due to spinal cord paralysis.[2-4] It is accepted practice to provide an aggressive pulmonary program for those patients developing acute respiratory failure due to neuromuscular disease. This includes endotracheal intubation or tracheostomy and mechanical ventilation.[5-6] The success of withdrawing or weaning ALS patients from artificial ventilation has varied from full pulmonary recovery to death.

Case Report

This case presentation describes a patient who was unable to be weaned from a respirator. Prolonged hospitalization for artificial ventilation was avoided when he was discharged home on a volume cycle (Bennett MA-I) respirator. It was felt that the medical care needed could be provided in the home setting with several possible advantages. Home management would: (1) decrease the risk of developing a nosocomial infection, (2) improve the patient's emotional outlook, (3) reassure the patient of having reliable and constant nursing care, and (4) decrease the cost of the medical care. The volume cycle respirator has been used in a home setting in a few patients, showing a marked decrease in the number of hospital days per year.[7] The real success of artificial ventilation at home is quite dependent on proper patient selection and the institution of a thorough home care program.

R.M. is a 37-year-old former Air Force navigator and a real estate broker. He first noticed in 1973 that he was unable to walk on his right heel. A few months later he developed a right foot drop, and sought medical attention. He underwent an extensive medical work-up as an outpatient at a university center over a six-month period. He was finally discharged on diet therapy, with the diag-

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nosis of mild chemical diabetes and amyotrophic lateral sclerosis, based on the clinical findings, electromyography, and progression of the disease.

R.M. returned to work and his normal daily routine, until he retired in August 1974, because he was unable to climb stairs and walk without crutches. In the summer of 1976 he was admitted to another hospital for an acute myocardial infarction, and was discharged on dipyridamole (Persantine) and clofibrate (Atromid-S). In January 1977 his wife divorced him. This forced him to move in with his parents because he was unable to care for himself.

R.M. was first admitted to the Bryn Mawr Hospital in August 1977 for tracheobronchitis with copious pulmonary secretions, which he was unable to handle because of an inadequate cough. Physical examination showed a chronically ill man who was using his accessory respiratory muscles, breathing at a rate of 32 breaths per minute. The blood pressure was 90/70 mmHg and the temperature was 100 F rectally. Examination of the chest showed no diaphragmatic movement, accentuated breath sounds, and a sinus tachycardia. Neurological examination showed some weakness of the lower jaw and sternocleidomastoid muscles. Speech was normal. There was generalized gross atrophy in all muscle groups both proximal and distal with fasciculations. There was also gross weakness both proximally and distally, such that he was only able to move his arms a few inches from the bed. There was no movement of the toes with plantar stimulation or with the Oppenheim maneuver. Chest x-ray films showed a poor inspiratory effort, but no acute parenchymal disease. The first set of arterial blood gases were on 2 liters of nasal oxygen and the results were: pH, 7.27; pCO_2, 67 vol %; pO_2, 102 vol %; and O_2 saturation, 97 vol %. His vital capacity was 700 cc. He was admitted to the Intensive Care Unit for pulmonary management.

The following day his arterial blood gases again on 2 liters of nasal oxygen were: pH, 7.28; pCO_2, 76 vol %; pO_2, 66 vol %; and O_2 saturation, 90 vol %. A tracheostomy was performed to better manage his secretions. Shortly thereafter, he became markedly fatigued and was intubated, and mechanical ventilation was started. After two weeks, his condition finally improved, and weaning from mechanical ventilation was started. All attempts at weaning the patient completely failed; respiratory fatigue was the limiting factor. His vital capacity remained less than 10 cc/kg and inspiratory force measured minus 15 cm H_2O.

R.M. became both physically and psychologically dependent on his MA-I respirator. The most R.M. was able to tolerate being off the respirator was for four hours with one hour on during the daytime, and continuous ventilation at night. The home use of the Bennett MA-I respirator was considered. It was felt that R.M. was an excellent candidate for home management because his pulmonary status had improved remarkably with minimal secretions and he could communicate with others by partially deflating the tracheal cuff, allowing air to pass through the larynx. Secondly, the location of the patient's home was only 15 minutes from the hospital by ambulance, and he could be easily transported in an emergency. Thirdly, R.M. was a very intelligent man and had an excellent understanding about both his disease and mechanical apparatus. Finally, his mother had volunteered to take care of him at home and learned tracheostomy care and pulmonary suctioning.

A home pulmonary program had to be planned. In the home there was a room adjacent to the kitchen which would be ideal for observing the patient. The room also allowed the patient to easily communicate with the members of the family.

The equipment needed at the home is shown in Table 1.

A pulmonary routine was set up around the needs of the patient and his family. Mrs. M. was available around the clock for tracheostomy care and suctioning as needed. A home health aide came into the home for four hours on Monday, Wednesday, and Friday, to give Mrs. M. relief. On Tuesday and Thursday a visiting nurse came to take vital signs and to give physical therapy and other nursing care. A respiratory therapist would visit twice a week for therapy and routine machine maintenance. His family physician would make a house call every two weeks for general medical evaluation of the program and to change the tracheostomy tube.

R.M. was discharged from the hospital on September 30, 1977. However, his initial stay at home was limited to two weeks, because his mother had to undergo cholecystectomy. He was again discharged on November 16, 1977. He enjoyed eight months at home, until he developed copious

Table 1. Equipment Needed for Home Ventilatory Care

1. Bennet MA-I respirator
2. A suction machine
3. An electric hospital bed
4. Mechanical hoist
5. Wheel chair
6. A loud call buzzer at the patient's bedside for emergencies
7. A back-up electrical outlet, for an emergency generator, which the local fire company would bring in a power failure
8. An adequate supply of tracheostomy tubes, sterile gloves, gauze, etc

amounts of sterile secretions which required suctioning throughout the night. He was again admitted to the hospital, and his secretions were managed by decreasing the humidity of the cascade at night. However, this hospitalization was complicated by congestive heart failure and diabetes mellitus. He slowly responded to diet changes, diuretics, and oral hypoglycemic agents. The patient was again discharged on September 15, 1978, and has remained at home to the present.

Discussion

The success of this patient's initial stay at home was unfortunately limited by his mother's illness. However, on his second attempt at home, he tolerated being on the MA-I respirator quite well for eight months. Hospitalization at that time was unavoidable because of the development of congestive heart failure and diabetes mellitus, which have a higher incidence in patients with ALS.[8,9] He again returned home and has been doing well.

It appears that the home use of the MA-I respirator had a definite benefit on the health and well being of this patient. Asmundson et al reported the incidence of acute necrotizing pneumonia in respiratory failure to be 13 percent, in which Pseudomonas, Klebsiella, and Proteus were the chief pathogens.[10] It is believed that these common hospital pathogens can easily be passed from patient to patient, by the common use of improperly cleaned respiratory equipment. In a home setting the patient is away from the hostile environment of the hospital, and he has the sole use of his own respiratory equipment.

The nursing care in the home did have some benefit over hospital nursing care. The care was adequate for outpatient needs as compared with the well-trained nursing care in the hospital. In the home there was always one person available to attend to the needs of the patient, and caretakers were limited to three or four different people throughout the year. In a hospital setting the care is usually not on an individual basis, and the personnel vary from day to day.

There were also definite emotional and psychological advantages to the patient when he was at home rather than in the hospital. He had become very confident in the care he was receiving at home. There was great apprehension in the hospital, when a new set of nurses or therapist took

Table 2. Rental and Service Fees per Month

Bennett MA-I respirator, including respirator therapy service and tubing change	$724
Electric hospital bed	80
Wheel chair	40
Hoist	35
Suction machine	30
Home health aid	451
Visiting nurse	360
Physician	60

over his care. Clinically, the patient was quite depressed in the hospital, for there was a loss of his feeling of privacy and independence. There was also a loss of time that was available to him to spend both with his parents and especially with his children on a weekend visit.

Finally, care at home on a respirator can be a large financial saving to the patient. The average cost of hospitalization on a respirator is approximately $9,000 per month. Home care is less than $2,000 per month (Table 2).

The cost of home respiratory care can be further reduced by purchasing certain pieces of equipment instead of renting them, especially when home care will be extended over long periods of time. Also, the home care can be reduced by the local chapters of ALS Foundation, Multiple Sclerosis Society, and Muscular Dystrophy Association which are providing patients with financial help for appliances and nursing care.

The use of an MA-I respirator at home is not ideal for all patients. Patients with respiratory failure from neuromuscular disease appear to be more appropriate candidates for home management than patients with intrinsic lung diseases. Patients with neuromuscular disease tend to be younger in age, and their disease is not usually complicated with primary lung disease or cardiovascular diseases. All patients should be screened to see if they are mentally and emotionally competent for home care. They should be able to communicate with others, and to tolerate brief periods of time off the respirator. The home pulmonary program should be formed around the needs of the patient and his family. There should be constant contact with the professionals involved, along with adequate medical records on the patient's progress.

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