

CASE REPORT

LONG TERM HOME VENTILATORY SUPPORT IN A PATIENT OF MOTOR NEURON DISEASE (MND) - A CASE REPORT

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ABSTRACT

A 45 years old male patient was diagnosed as a case of motor neuron disease. Within four years he became paralyzed and bed ridden. Due to severe respiratory tract infection, the patient developed respiratory failure and was put on ventilator. Later on the infection was successfully treated but the patient could not be weaned from the ventilator because of the paralysis of respiratory muscles. It was obvious that the patient would require life long ventilatory support so the same was arranged at the patient's home and he was discharged from the hospital after nine months. Now, for the past six years the patient is on ventilatory support.

KEYWORDS: Motor Neuron Disease, Respiratory Failure, Life-long Home Ventilatory Support.

INTRODUCTION

Motor neuron disease is a premature ageing process resulting into degeneration of upper as well as lower motor neurons of spinal cord and brainstem. There is slow, progressive muscular weakness leading to paralysis of almost all voluntary muscles. Respiratory failure is the end result when the degenerative process involves the respiratory muscles. Since long-term ventilation is a major undertaking the decision is made after extensive liaison with patients, the family and numerous specialized support services. The patient and his family members are then educated on care of the system at home.

CASE REPORT

The history of the patient dates back to about twelve years when a 45 years old male patient of

good build presented with weakness of right big toe. The weakness slowly progressed to involve both lower limbs. He was diagnosed as a case of motor neuron disease. As the patient was in USA at that time, he was initially treated there, and he got membership of American Society of motor neuron disease. Later on the patient returned to Pakistan. Six years back he developed severe respiratory tract infection and respiratory failure. The patient was admitted in Intensive Care Unit of Combind Military Hospital, Lahore and placed on ventilatory support. Keeping in view the need of long term ventilation, tracheostomy was performed and ventilatory support continued through tracheostomy tube. His throat and blood cultures yielded staphylococcus and pseudomonas, sensitive to cephalosporins. For three to four days patient was kept sedated and parenteral antibiotics and nasogastric tube nutrition were started. Necessary monitoring was ensured e.g. recording of pulse, Blood Pressure, ECG, pulse oximetry, capnography, arterial blood gases, urine output and central venous pressure.

After two weeks, the patient became afebrile

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and blood culture negative. Then the weaning process was started with Synchronized Intermittent Mandatory Ventilation (SIMV) with pressure support. All efforts were made to wean him off the ventilator but the patient was unable to sustain due to involvement of his respiratory muscles. As the patient started swallowing, nasogastric tubing was removed.

The patient was educated about the posture in the bed, the method of diaphragmatic breathing and weaning exercises. Also the confidence building measures were adopted to alleviate his fear of impending death. Recreational facilities were provided to the patient like a television. Provision of light and dark hours was ensured to keep the diurnal rhythm intact. During night the patient was provided with full ventilatory support but in daytime it was kept intermittent or assisted as per patient's requirement.

Patient adapted well to the process of artificial respiration. He had good orientation and could communicate by writing. Within next two months, patient was found to be so comfortable with ventilator that all invasive monitoring was stopped and intravenous canulae removed. After 9 months of his stay in the hospital the patient was shifted home continuing his ventilatory support. Extensive discussions were made with close relatives and the patient himself and they were educated on day-to-day management of the system. Two ventilators (Adult Star®), suction apparatus and central oxygen supply were fixed in the patient's bedroom.

Now for the past six years the patient is doing well with ambulatory ventilatory support at home. Most of the time he is ventilated on air but according to physical needs 30% oxygen is occasionally added. Patient requires frequent suction that is done by his attendants. A portable X-ray machine is provided at home accordingly. Patient can sit in the lawn for 2-3 hours and have chat with friends and family members. Outside, someone ventilates him with help of an Ambu® bag. Similarly, he can also travel on the back seat of his

car for 2-3 hours along with an attendant with assisted Ambu® bag ventilation. Slowly the degenerative process has involved patient's upper limbs also and thus his writing is also becoming jeopardized.

Initial cost was about Rs. 5.5 million and the daily cost is about Rs. 1000 to 2000.

DISCUSSION

Motor neuron disease is a relentlessly progressive disorder leading to respiratory paralysis. The disease affects both upper and lower motor neurons. On the basis of predominance, its chronic form is sub-grouped as amyotrophic lateral sclerosis (ALS), bulbar or pseudobulbar palsy, spinal muscle atrophy or primary lateral sclerosis and other patterns. As the muscles are denervated, there is progressive atrophy of muscle fibers but, remarkably, sensory neurons, autonomic function, coordination and higher cerebral function are spared.¹

There are rare reports of stabilization or even regression of ALS. While ALS is overwhelmingly a sporadic disorder, some five to ten percent cases are inherited as an autosomal dominant traits. In most societies there is an incidence of 1 to 3 per 100,000 and prevalence of 3 to 5 per 100,000. Males are affected more than females.²

The precise cause of MND is unknown. Postulated pathogenic causes include oxygen free radicals, excessive excitatory neurotransmitters, growth factor and immunological abnormalities.³

Clinically any muscle group may be first to show signs of disease, but, as the time passes, more and more muscles becomes involved and ultimately the disorder takes on a symmetrical distribution in all regions. Mostly in later stages of illness sensory, bowel and bladder and cognitive functions are preserved. Dementia is not a component of disease, especially ALS.

Diagnosis is almost always made on clinically grounds together with electromyographic (EMG)

evidence of denervation. Differential diagnoses are cervical cord or brain stem compression, chronic heavy metal poisoning and chronic progression of poliomyelitis (post polio syndrome).⁴

Death mostly occurs due to respiratory infection, aspiration or respiratory failure or ventilatory failure from prolonged weakness and paralysis.

Management is symptomatic and supportive. Admission is required with acute progression of disease. The intensivist may be asked for ambulatory or home ventilatory support for gradually worsening chronic respiratory failure. These patients present major ethical and clinical problems. Since positive pressure ventilation by mouth or nose provides transient relief, long-term ventilation by tracheostomy is life sustaining. Provision of Continuous Positive Airway Pressure (CPAP), pressure support or other modes of assisted ventilation can result in an improved quality of life and possibly prolonged survival for carefully selected individuals.⁵

Some patients require only intermittent support, particularly at night or during periods of acute deterioration due to intercurrent illness. Weaning exercises should be started as early as possible but in such cases weaning from mechanical ventilation is mostly a game of patience and can rarely be achieved with success. Special care is warranted to avoid respiratory infection and other complications associated with prolonged immobility.

Long-term respiratory or ventilatory support outside ICU is a major undertaking, requiring specific equipment and excessive liaison with the patient, the family and multiple specialized support services.⁶

Looking retrospectively at this particular case being sustained on artificial means of life support for more than six years, this sort of endeavor is worth practicing.

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