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Breathing Pacemaker in Poliomyelitis -A Case Report.

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Abstract

Poliomyelitis is a disease of ambiguity that usually affects the children. Though the disease commonly affect the lower limbs, rapidly developed quadriplegia with progressive respiratory difficulties are often encountered. It can be diagnosed confidently with Physical examination and stool culture. Such a quadriplegic case with respiratory drive failure is resuscitated with ventilator in intensive care unit. Rehabilitation with breathing pacemaker system is an acceptable way of weaning the ventilator. Advantage of breathing pacemaker system over the ventilator is numerous. The system was made available and a team of surgeons operated to implant it. Externally placed transmitter and antennas transmit energy to the bilaterally implanted receivers, which convert the radio signals into stimulating pulses delivered to both phrenic nerves by the corresponding implanted electrodes. The hemidiaphragm then contract, producing inhalation of air. The train of pulses then stops, allowing the diaphragm to relax and exhalation to occur in a passive process. This cyclic repetition of pulse trains followed by no pulses produces a normal breathing pattern. The total system can be monitored through a system via telephone called "Trans Telephone Monitoring" (TTM). The diaphragm, like other skeletal muscles when denervated is expected to atrophy and therefore, become fatigued upon stimulation. Re-training and reconditioning programs are being carried out to have the full functioning of the pacemaker. The boy now improving very slowly and is discharged with a portable ventilator. This case note represents the first case of poliomyelitis with breathing pacemaker so far.

Key Words: Poliomyelitis, Breathing Pacemaker, TTM, Ventilator.

Introduction

Cavallo first proposed the use of electrical stimulation to artificially controlled respiration in 1777 shortly after the discovery of electricity. More conventionally electrical stimulation has been used to provide clinical ventiltory support for ventilator dependent quadriplegics, patients with central hypoventilation syndrome and patients with sleep apnoea. The external radio frequency transmitter sends signals through two antennae taped to the skin. These stimuli are picked up by bilateral receivers implanted under the skin and electrical pulses are delivered by electrodes attached to the phrenic nerves. Bursts of pulses delivered to the phrenic nerves

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cause each hemidiaphgragm to contract giving rise to expansion of the lungs (inhalation of air). When the train of pulses stops, the diaphgragm relaxes and exhalation occurs in the passive way. These bursts of pulses are typically applied for 1.6 seconds and repeated 12 times per min. permitting 24 hours breathing in quadriplegics without a tracheostomy. Breathing pacemakers have been used to treat patients with phrenic nerve damage, multiple sclerosis, GBS, obstructive pulmonary disease, spinal muscular atrophy, idiopathic diaphragmatic paralysis and chronic hiccup[2-5]. Data of 700 patients ranging from younger than 2 months to 89 years of age demonstrate that some of these patients have been successfully undergoing pacing for more than 20years[6].The limiting factor is whether the phrenic nerve and the diaphragm muscles are intact or whether they can be stimulated with suitable current stimulus. Breathing pacemaker in polio patients are not known. This case report demonstrates the first implanted breathing pacemaker in poliomyelitis.

The Case Report

Amit, 10 years old school boy from Dhaka city was admitted in the paediatric unit of Bangabandhu Sheikh Mujib medical university hospital, Dhaka, during september 1999. He had complaints of mild continuous fever, headache and pain in the limbs for 5 days. Following the 2nd day of hospital admission, headache was further intensified with addition of his neck stiffness and weakness in the limbs. Within next few hours, limb weakness further deteriorated and he developed frank quadriplegia with rapid respiratory insufficiency. General and neurological examination revealed, normal higher psychic function and no motor function in the limbs, cranial nerve function was normal. Deep tendon reflexes were absent. There was mild dissociation of pain, touch and temperature sensations below C2 on the right side and C3 on the left. Routine laboratory investigations revealed no abnormality. Lumber puncture demonstrated significantly raised CSF protein, reduced sugar and a normal electrolyte content with sterile culture. Stool sample was sent for poliovirus. He was given a course of interferon with the differential diagnosis of GBS, and poliomyelitis. Following further deterioration of respiratory function he was transferred to intensive care unit. An emergency tracheostomy was done and he was put to a ventilator and PPMV was continued. Stool report confirmed polio with isolation of P -1 virus.

After 10months in ICU, he was presented in a clinical meeting, where suggestions are sought from different specialists regarding further management. Rehabilitation team proposed for the possibility of a breathing pacemaker.

Clinical examination during August 2000 revealed, he was full conscious and oriented. All 4 limbs were wasted grossly with just flickering of muscle movements. Level of sensori-motor quadriplegia was below C3. Deep tendon reflexes were absent. Routine chest and pulmonary function tests demonstrated that he has a forced tidal volume of 0.35L with some reduced function on the left lung and a normal functioning right lung. Transcutaneous stimulation of the phrenic nerve in the neck was not successful. EMG of intercostal muscles showed positive sharp waves with increased insertional activity. EMG of the diaphragm and the phrenic nerve conduction velocity was not possible due to technical difficulties. It is assumed that he is going to be permanently ventilator dependent and a partial functioning of the phrenic nerves are possible. Breathing pacemaker is an alternative to ventilator, which works in partial functioning phrenic nerves as well, is much superior physiologically and is

cost effective. The viability of the phrenic nerves was determined intra-operatively during implantation operation under direct vision.

Breathing pacemaker system was made available. It costs about 76,000 US dollar. A team of surgeons with the-onsite instructions of a biomedical engineer, operated to implant the system using the standard fascian of surgery. Testing with sterile antenna prior to closure demonstrated contractions of both hemi diaphragms confirming the integrity of the phrenic nerves. Post operative come-up was un-eventful.

After 4—weeks of implantation operation, a standard protocol was begun to stimulate the diaphragm. Initially, the patient was given 3 minutes of pacing. Within the course of a month, he was able to tolerate 10 minutes of pacing of the diaphragm with constant complaints of tiredness, sweating and tachycardia. Diaphragm movement was confirmed by fluoroscopic examination. "Trams Telephone Monitoring"(TTM) was not possible due to technical difficulty. Over the period of next 3 months, the patient showed slow improvement of pacing time. He was discharged with a portable ventilator. Father of the child was trained to operate the portable ventilator with on call assistance of an ICU doctor. At the end of one year of implantation operation, he could continue respiration for 4 hours per day using the pacemaker system.

Discussion

The diaphragm is the principal muscle of respiration. Motor supply of the diaphragm is by the phrenic nerve whose root value is from C3 to C5, mainly C4. In an individual with a spinal cord lesion above C3, the diaphragm becomes paralyzed but the phrenic nerve is expected to be viable. Poliomyelitis affects the anterior horn cells of the spinal cord or motor nuclei of the brain. A small fraction of the patients with paralytic polio need chronic respirator care. The return of muscle strength occurs mainly in the first 3-4 months and is the result of enlargement of motor units by re-innervation and by morphologic restitution of partially damaged nerve cells. Slow recovery of slight degree may then continue for a year or more. Branching of axons of intact motor cells with re-innervation of muscle fibres of denervated motor units may also play a part[7].

Transcutaneous stimulation of the phrenic nerve was found to elicit response of the diaphragm in 1984[8] and since then electrophrenic respiration was applied. Electrophrenic respiration through implanted radio-frequency stimulator was first introduced by van Heekren and Glenn in 1966[9] First pacing was used in patients with Ondyne's curse. With this initial success, breathing pacemaker found broader application in ventilator-dependent patients with respiratory drive failure. Glenn and co-workers reported the first case of total ventilatory support of a C2 quadriplegic[10]. All the above conditions require the phrenic nerve to be intact to allow the conduction of the artificial stimulus to the diaphragm. Axonal continuity should be tested before implantation of a pacing system[11] although we could not make the facilities available in our case. Transcutaneous stimulation of the phrenic nerve has been shown to be successful in 95 of 100 patients[11] however, failure of the diaphragm to contract when transcutaneous stimulation is applied to the phrenic nerve does not necessarily mean that the nerve will not respond to direct stimulation. Definitive test of phrenic function is achieved via open dissection of the phrenic nerve, as happened to our case. TTM feedback is an useful way of monitoring the system but due to technical difficulties we could

not avail it.

The denervated muscle is expected to atrophy and, therefore, become quickly fatigued upon stimulation. So it requires a reconditioning period that may take a period even of two years or more.

Conclusion

This case represents the first implanted breathing pacemaker in polio patients. Diaphragm pacing provides physiological respiratory function far superior to that provided by mechanical ventilators since the inhaled air is drawn in, rather than being forced into the chest under mechanical pressure. The benefit of breathing pacemaker over the ventilator is numerous. 24 hours continuous pacing is expected although the presented case is improving very slowly.

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