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Effect of Breathing Exercise on Improvement of Pulmonary Function in Patient With Amyotrophic Lateral Sclerosis: Case Study

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국문요약

근위축성 측색 경화증 환자에서 호흡운동 치료가 폐기능에 미치는 효과

정영종
국립암센터 물리치료실

근위축성 측색 경화증 (amyotrophic lateral sclerosis: ALS) 환자에게 있어 호흡기능 장애는 죽음에 이르게 하는 주요 원인 중 하나이다. 본 연구는 근위축성 측색 경화증이 있으며 호흡기능이 약화되어 있는 51세의 여성 환자를 대상으로 호흡운동 치료를 시행한 후 폐기능(pulmonary function)이 증진되었는지를 알아보고자 실시하였다. 연구 대상자는 6주간의 호흡운동 치료 프로그램에 참여하였다. 호흡운동 치료 프로그램은 횡경막 호흡(diaphragmatic breathing), 복부근육강화(abdominal muscles strengthening), 지갑입술 호흡(pursed lip breathing), 그리고 동기 유발성 흡기폐활량계(incentive spirometer)를 이용한 흡기운동 등으로 구성되었다.

폐기능 검사는 이동식 호흡측정기(spirometer: MICROSPIROHI-198)를 이용해서 시행하였다. 또한 하지 에르고미터(cycle-ergometer)를 이용해 운동 시간을 측정함으로써 폐기능의 증진 여부를 알아보았다. 연구 대상자는 6주간의 호흡운동 치료 기간 동안 노력성 폐활량(forced vital capacity: FVC)과 정상 예측치에 대한 노력성 폐활량의 비율(percentage of the predicted forced vital capacity: %FVC), 그리고 하지 에르고미터의 운동 시간에 있어 현저한 증가를 보였다. 그러나 노력성 폐활량에 대한 1초간 노력성 폐활량 비(FEV1/FVC)에 있어서는 약간의 감소를 보였다.

근위축성 측색 경화증 환자에게 6주간의 호흡운동 치료를 실시한 결과, 폐기능의 증진에 효과적임을 알 수 있었으며, 앞으로 더 많은 연구 대상자에게 그 효과를 알아보는 연구가 필요할 것이다.

핵심단어: 근위축성 측색 경화증; 폐기능; 호흡운동치료.

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive, neurodegenerative disorder of the voluntary motor system that is characterized by loss and degeneration of motor neurons and their outflow tracts (Kaplan and Hollander, 1994). ALS primarily affects the motoneurons of the cerebral cortex, brain stem, and spinal cord (Brown, 1997;

Hirano, 1996). Involvement of the motor neurons for the respiratory muscles eventually leads to dyspnea and then respiratory failure, the most common cause of death in ALS (Caroscio et al, 1987; Tandon and Bradley, 1985). The incidence of ALS is .4 to 2.4 cases per 100,000 people worldwide, with a prevalence of 2.5 to 7 cases per 100,000 people (Juergens and Kurland, 1980; Mitsumoto et al, 1998; Tandon and Bradley, 1985).

No cure exists for ALS, but medications have beneficial effects. Riluzol (Rilutec) inhibits glutamate release and antagonizes the glutamate receptor, which prolongs survival (Bensimon et al, 1994).

The course of the disease is inexorably progressive (Arnulf et al, 2000). During the course of the disease, the involvement of various respiratory muscle groups leads to a restrictive ventilatory defect and ultimately to hypercapnic respiratory insufficiency (Arnulf et al, 2000). In the majority of cases, death is related to respiratory events (Caroscio et al, 1987; Tandan and bradley, 1985). Among the mechanisms of respiratory impairment in ALS, diaphragmatic dysfunction probably plays an important role (Caroscio et al, 1987). Attali et al (1997) reported that diaphragmatic dysfunction, defined as the presence of abdominal paradox and delayed or abolished responses of the diaphragm to phrenic nerve or cortical stimulation, was generally associated with dyspnea. Patients with ALS usually die from such respiratory problems as hypo-ventilation causing hypoxemia, hypercarbia, or both; aspiration pneumonitis; other pneu-monia; and occasionally pulmonary embolism (Caroscio et al, 1987; Kaplan and Hollander, 1994). It has been recently reported that patients with ALS exhibited features typical of diaphragm paralysis with compensatory use of inspiratory neck muscles, and It was suggested that diaphragmatic dys-function is a major determinant of dyspnea in ALS (Attali et al, 1997; Similowski et al, 1999).

Respiratory symptoms usually appear late, and the monitoring of lung and respiratory muscle function have been reported to be the best prognostic indicators in these patients (Fallat et al, 1979; Kaplan and Hollander, 1994; Schiffman and Beish, 1993).

Nakato et al (1976) were among the first to report serial pulmonary function studies in ALS. They measured lung volumes and diffusing capacity, but not static pressures, in 25 patients beginning 9 months after onset and at 15, 22, and 33 months. Initial mean vital capacity (VC) was within the normal range and averaged 58 percent of predicted at final measurement. Schiffman and Beish (1993) reported that 31 of 36 patients had respiratory muscle weakness at presentation, although only 7 complained of any respiratory symptoms. Vital capacity (percent predicted) was significantly lower in the symptomatic group (55.9) compared with the asymptomatic group (76.4). It is concluded that early measurement or respiratory muscle strength in ALS with subsequent follow-up studies may be useful in determining overall prognosis and in decision making.

For many years, physical therapists have instructed patients in breathing exercise.

Breathing involves movement of many joints and contraction and relaxation of many muscles. It is a complex movement that happens most of the time without consciousness. The purpose of breathing exercises is to reduce the work of breathing and to improve the ventilation and perfusion of the lungs. As the majority of respiratory function is weakened in ALS, breathing exercise and education is very important.

To improve respiratory capacity, I chose the diaphragmatic breathing control, pulsed lip breathing, thoracic mobility, incentive spirometer, and especially strengthening of the abdominal muscle during inspiration.

Fallat et al (1979) reported the use of spirometry (but not static pressures) at the time of diagnosis and serially in a large number of patients with motor neuron disease, mostly with ALS. At the time of diagnosis, 93.6% of their patients showed at least one abnormality for the following three parameters: forced vital capacity (FVC), maximum voluntary ventilation (MVV), and residual volume (RV). Munsat et al (1988) have reported a linear loss of muscle function, including pulmonary function, during the “active phase of the illness” in ALS patients. Early in the disease, before an area of motor function such as a limb becomes involved, there is a period of stability. Once an area of the body starts to weaken, the decline in strength over time is linear until the low end of function is reached and the plot levels out without further decline. Although rates of decline vary among patients, the plots tend to be linear in individual patients. FVC was the only measure of pulmonary function in this study, and its plot over time was also found to be linear (Andres et al, 1986; Munsat et al, 1988).

The abdominal muscles are the primary muscles of expiration. They also have an important role in augmenting inspiration in the upright posture (Danon et al, 1979; De Troyer, 1983). De Troyer (1983) proposed several mechanisms by which abdominal muscle recruitment may confer mechanical advantage to the diaphragm. Abdominal muscle activity and tone prevent excessive shortening of the diaphragm after each contraction, enabling the diaphragm to function on a more favorable position of its length-tension curve. The increase in abdominal pressure developed during inspiration is transmitted to the lower rib cage. During exercise, abdominal muscle activity is even more important. Abdominal muscle recruitment for active expiration begins at low levels of exercise when minute ventilation exceeds 30 L (Pardy et al, 1984). The tidal volume increase during exercise is in both the inspiratory and the expiratory directions. The decrease in functional residual capacity (FRC) is preferentially through the abdominal pathway (Grimby, 1968). Displacement of abdominal contents upon contraction of the abdominal musculature pushes the diaphragm upwards so that it operates on a more favorable portion of its length-tension curve during inspiration. Dodd and colleagues (1984) emphasized another mechanism for the role of abdominal muscles as accessory muscles of inspiration. They suggested that the recruitment of the abdominal muscles during exercise leads to a storage of elastic and gravitational energy, which, when released

during inspiration, contributes to generation of negative pleural pressures and enhanced inspiratory flow (Dodd et al, 1984).

Although respiratory function plays such an important role in the prognosis of these patients, there have been few reports of respiratory function studies in ALS (Black and Hyatt, 1971; Keltz, 1965; Nakano et al, 1976; O'Donohue et al, 1976).

As the majority of patients presenting with ALS already have evidence of respiratory muscle weakness, I think that it may be useful to establish a baseline with the effect of pulmonary rehabilitation. Pulmonary rehabilitation is an intervention that can combine exercise, education, and behavior modification strategies in an effort to minimize symptoms and to improve quality of life (Camp et al, 2000).

Case Study

History

A 51-year-old female suddenly experienced whole body weakness, followed by fatigue especially in the afternoon approximately 7 years ago. Her weight was 58 ㎏, and height was 164 ㎝.

Since then, the patient had received some medical treatments and physical therapy, but was not informed of the exact diagnosis. After 2 years, the diagnosis of ALS established in the patient by clinical examination, electromyographic studies, nerve conduction studies, and further neurologic procedures that were necessary to exclude other neuromuscular disorders. The patient history included an both extremities paralysis and some sleep disturbance. After paralysis, the patient felt extreme weakness, had difficulty in walking and active daily living activity. After the hospitalization the course of the disease is inexorably progressive. The patient complained of feeling fatigued and being uncapable of singing a song due to shortness of breath, and having difficulty in coughing and deep breathing. Especially the patient could not perform the lower extremity ergometer for more than 2 minutes.

The patient lacked knowledge of ALS, its effect on respiration, methods to control dyspnea, breathing and coughing techniques, and methods of controlling respiration during exercise. All of these symptoms had been unchanged for 6 months prior to referral to breathing rehabilitation program. The patient and her family were informed of the diagnosis and the progressive nature of the disease. I believe it is important that the patient and family be given as much information as possible about the disease process, so that informed decisions about care can be made. The patient participated in goal setting and treatment planning.

Evaluation

Respiratory function test was performed at 6 pm, twice a week, every Tuesday and Friday. Respiratory function was tested by a portable spirometer (MICROSPIRO HI-198). Spirometry is considered of value, not only in detecting early respiratory involvement in ALS, but in detecting early respiratory involvement in ALS, but also in predicting the course of respiratory failure (Kaplan and Hollander, 1994; Ringel et al, 1993). Spirometry was performed with the patient wearing a noseclip and connected directly

by a mouthpiece. The data was digitally recorded. The test was performed 3 times to meet reliability guidelines and the measurements of the best FVC of three trials were recorded, not necessarily from the same trial. Resting period of five minute were provided to allow ample recovery from the fatigue. After the final testing session, the patient was participated in the interview. The initial respiratory function evaluation was performed for 10 days as a baseline information.

Treatment

Patients were given detailed education covering the following: breathing and coughing techniques, methods of control of dyspnea, signs of illness and time to seek medical help, energy conservation techniques, nutritional and exercise guidelines, and body mechanics. Teaching the patient and his family the indications for and the importance of his treatment decreases anxiety and increases compliance. These are vital points to consider if maximal treatment benefit is to be realized and further complications avoided.

The main objective of the pulmonary rehabilitation program was to promote respiratory function by means of breathing exercises. Pulmonary rehabilitation management also included education about self-monitoring and progression of activity. Physical therapy began 2 weeks post-admission and consisted of breathing exercises. Three times a day physical therapy session (9 am, 1 pm, 5 pm), duration of 20 minute were planned for the patient, 5 days a week, for a 6-week period. If the patient had been fatigued, the patient was not treated by the physical therapist. The frequency and duration of the exercise prescribed depend on the patient's level of exercise capacity. Repetitions of exercises were progressively increased as the patient's dyspnea improved and the patient was more able to tolerate the pulmonary rehabilitation program. During this time, The patient had a 4 minute warm-up and cool-down phases. The warm-up and cool-down phases consisted of stretching exercises for upper and lower extremities, thoracic mobility exercises.

Wong (2000) reported that the upright sitting position was considered the position of choice for intervention because the patient's arousal was at its greatest, ventilation-perfusion ratio was optimal, and diaphragm muscle excursion was maximal in an upright position. In this position, the patient performed the breathing exercises. Head and upper body was leaned against the wall very relaxed position. The work of breathing was also reduced in that position. The active cycle of breathing techniques was incorporated with periods of breathing control, deep breathing mobilization/exercise, and cough. During the breathing control period, the patient was instructed to perform normal tidal breathing using the lower chest wall and to relax his upper chest and shoulders. Verbal input and tactile input (ie, the therapist's hand was placed over the patient's abdomen) were given until the patient was able to achieve breathing control. During deep breathing, the patient was instructed to inhale deeply through the nose (ie, slowly over 3 5

seconds) to inspiratory reserve volume (when he began using his accessory muscles) and to exhale through the mouth passively. The emphasis was on lower chest wall expansion. There were four repetitions of deep breathing followed by a period of breathing control. Before breathing exercises were performed, mobilization and exercise in the form of arm elevation and thoracic mobility (rotation) exercises (10 20 times or as tolerated) were prescribed to optimize the various steps of the oxygen transport system, particularly to stimulate maximal inspiration and facilitate mucociliary transport rate.

For improving of respiratory function, the patient performed the inspiratory exercise using inspirator, abdominal muscle strengthening exercise using manual contact, and diaphragmatic breathing exercise with pursed lip breathing technique. Inspiratory muscle training and resistive diaphragmatic breathing exercises may be beneficial while weaning the patient with quadriplegia or chronic pulmonary disease form the ventilator (Aldrich and Karpel, 1987; Derrickson et al, 1992; Hornstein and Ledsome, 1986). Diaphragmatic breathing exercises increase lung volume and improve gas exchange (Levenson, 1992). Exercise training of inspiratory muscles may be indicated to improve their function (Reid and Dechman, 1995). Energy conservation techniques can be useful in helping a patient accomplish daily activities with less effort expended and to spread the effort throughout the day (Ringel et al, 1993).

Strengthening of the abdominal muscle has been described as one of the major aims of therapy for respiratory function. Abdominal muscles are the principal muscles of active expiration, and cough is largely (Tandan and Bradley, 1985), though not exclusively (Bensimon et al, 1994), dependent on the integrity of this muscle group. Strengthening exercise for the abdominal muscle was to be performed everyday. I chose this treatment approach because I believed the patient did not use abdominal muscles selectively during breathing. I thought this problem was one of the respiratory failure.

Inspiratory exercise using spirometer was performed in comfortable sitting position.

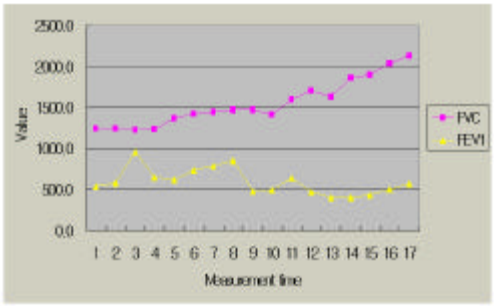

Fig 1. Spirometric testing results (FVC, FEV1)

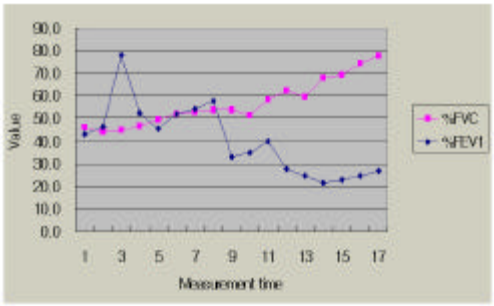

Fig 2. Spirometric testing results (% FVC, % FEV1)

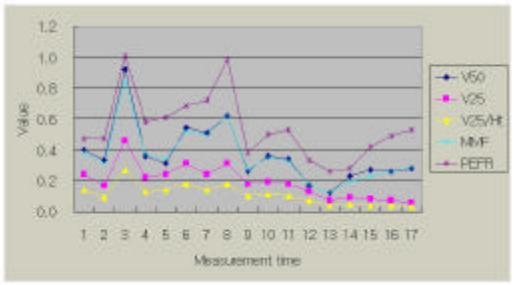

Fig 3. Spirometric testing results (V50, V25, V25/Ht, MMF, PEFR)

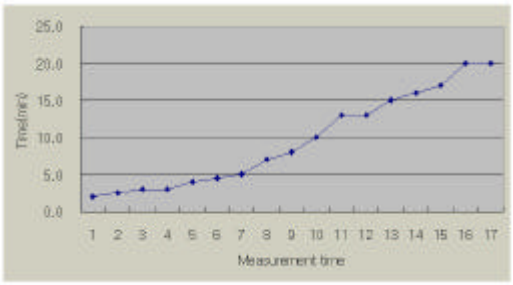

Fig 4. Ergometer exercise time

Results

Spirometirc data were illustrated graphically (Fig 1-3). Figures showed that the effect of the pulmonary rehabilitation program in ALS. Initially, the patient showed severe restrictive pattern as assessed by a FVC that was lower than 50% predicted value. There was a definite progression to a lower FVC with 6 consecutive weeks of the pulmonary rehabilitation program from 1230 to 2130 cc. Three weeks after the pulmonary rehabilitation program, FVC was increased dramatically.

At initial evaluation before pulmonary rehabilitation program, she was able to perform the lower-extremity ergometer for approximately 3 minutes, however the duration of exercising gradually increased to 20 minutes (Fig 4).

Discussion

Pulmonary rehabilitation program, as given in this study, was able to improve quality of life of the patient by delivering exercise training, education, and coping strategy techniques. Despite the poor prognosis of ALS, I believe that pulmonary rehabilitation program is an important component of the overall care of patients with this disease. Through active participation in treatment planning, the patient may gain some sense of control over what is happening to her body and enable her to cope with functional losses efficiently. The major impact of pulmonary rehabilitation was improvement in quality of life.

Weakness of the respiratory muscle is an obligatory component of ALS. Moreover, progression in ALS is faster than in most other chronic neuromuscular disorders explaining why respiratory failure is the most common cause of death (Juergens and Kurland, 1980). Patients with amyotrophic lateral sclerosis with mild restrictive ventilatory pattern may show reduction in respiratory muscle function, rapid shallow breathing, and a preserved neural drive.

Respiratory function plays such an important role in the prognosis in ALS. Thus, pulmonary rehabilitation program involving breathing exercises is essential treatment in ALS. Since ventilatory function is so consistently affected in patients with ALS, I used the spirometry as a simple

and accurate means to assess the severity and the progression of the disease objectively. Although ALS is usually fatal, the time course of the symptoms is quite variable, as emphasized in previous reports (Kaplan and Hollander, 1994; Serisier et al. 1982). Impairment in FVC is a possible harbinger of a more rapid clinical deterioration.

Spirometric measurements show that in a majority of cases, regardless of the pattern of motor neuron impairment, FVC decreases with disease progression and is correlated with survival (Black and Hyatt, 1969; Fallat et al, 1979; Nakato et al, 1976; Schiffman and Belsch, 1989). The values for the patient was compared with her predicted normal value that was adjusted by age, sex, and height. Forced expired volume in 1 second (FEV1) and FVC are among the most reliable of spirometric measures. The decline of FVC is occurred in every disease affecting the bellow action of chest wall and expansion of lung. The low FEV1/FVC was related to her weak and uncoordinated expiratory effort. FEV1/FVC ratio of less than 0.6, which indicates a greater degree of chronic airflow limitation. The %FVC and %FEV1 were calculated by dividing the actual value by the predicted value for her. Mahler et al (1992) found that measures of pulmonary function (%FVC and %FEV1) were related to disability. The most consistently and significantly reduced measurement was the %FEV1. These findings provide objective confirmation in pulmonary function of the clinical impression during pulmonary rehabilitation program to her.

There were some limitations to this study. These limitations included the lack of subjects, the inability to determine the long-term benefits of pulmonary rehabilitation. More research on pulmonary rehabilitation program is needed if therapists are to make evidence-based decisions regarding the respiratory function. Randomized studies that compare the effects of different exercise programs in terms of type, frequency, duration, and intensity of exercises, for example, are needed. Outcome measures should include not only force measurements but more importantly, function and quality-of-life measures.

Physical and emotional health are affected by pulmonary rehabilitation in ALS. Because the patient was exuberant, alert, and cooperative, the patient could get more benefit from breathing exercises to improve tidal volume, thoracic-cage mobility, inspiratory capacity, and cough efficacy. Above all, the patient could find the pleasures in her life. I believe that physical therapist's care and compassion can have an impact on the well-being of people living with disease.

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