Benefit of Forced Expiratory Technique for Weak Cough in a Patient with Bulbar Onset Amyotrophic Lateral Sclerosis

MITSUAKI ISHII, RPT1)

1)Department of Physical Therapy, Maizuru Municipal Hospital:
150–11 Mizoshiri, Maizuru, Kyoto 625-0035, Japan.
TEL +81 773-62-2630  FAX +81 773-64-6200  E-mail: mm.ishii@orion.ocn.ne.jp

Abstract. The specific purpose of this case study was to investigate whether forced expiratory technique (FET) improves the peak expiratory flow compared to coughing in a 53-year-old man with amyotrophic lateral sclerosis (ALS) who presented with bulbar symptoms. Approximately 12 months after diagnosis, his peak cough flow did not exceed 160 L/min, and cough became ineffective. However, FET could generate peak expiratory flow to a point over the 160 L/min threshold until 14.5 months after diagnosis. As a result, FET delayed the need for tracheostomy. When the forced vital capacity (FVC) was observed to be markedly decreased and it was 1,600 mL, the patient was unable to achieve 160 L/min of peak expiratory flow generated by FET. Patients with bulbar onset ALS who have FVC greater than 1,600 mL may benefit from FET.

Key words: Amyotrophic lateral sclerose, Bulbar involvement, Forced expiratory technique

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterized by loss of motor neurons1). In approximately 25 percent of patients with ALS, the initial symptoms begin in muscles innervated by the lower brainstem cranial nerves in the bulb (medulla)2).

As bulbar muscle dysfunction progresses, it impairs the retention of optimal breath with a closed glottis, and cough becomes ineffective3, 4). During cough, high peak intrapulmonary pressures are reached when the glottis is closed, and with opening of the glottis, high expiratory flows are generated. Incomplete glottic closure can exacerbate cough dysfunction and further decrease peak cough flow (PCF)5). The inability to effectively cough is associated with aspiration or respiratory infection, and is a cause of respiratory failure and death.

The author hypothesized that use of the forced expiratory technique (FET)6) known as huffing, that creates forced expiratory flows through an open glottis, might be useful in the treatment of patients with ALS presenting with bulbar involvement. FET is a technique to assist in the expectoration of secretions. Unlike a cough in which the glottis is closed, FET requires the glottis to remain open. The technique is taught when cough is ineffective. It has been proposed that FET for patients with chronic bronchitis is an important adjunctive therapy7). However, little has been reported about the effect on FET in the patients with bulbar onset ALS. The essential purpose of this case study was to investigate whether a patient who had bulbar involvement but adequate other muscles function can benefit from FET compared with coughing on the peak expiratory flow.
CASE REPORT

The patient was a 53-year-old man who noticed dysphagia in November of 2002 and dysarthria in January of 2003. In May, he developed weight loss and was subsequently admitted to the University hospital for further examination and management. During this hospitalization, he was diagnosed with ALS and was prescribed Riluzole. He had no upper motor neuron signs. However, physical examination and an electromyographic study revealed that the patient had lower motor neuron signs in the bulbar and cervical regions. Based on progressive muscular weakness with fasciculation in at least two body regions, he fulfilled El-Escorial diagnostic criteria for “suspected ALS”.

Pulmonary function testing revealed 3,730 mL of vital capacity (VC), 102 percent of predicted VC, and 79.8 percent of forced expiratory volume in one second per forced vital capacity (FEV$_1$). Arterial blood gas values were pH, 7.445; arterial partial pressure of carbon dioxide (PaCO$_2$), 40.5 mmHg; partial pressure of oxygen (PaO$_2$), 97.8 mmHg; bicarbonate (HCO$_3^-$), 27.2 mEq/L; and base excess (BE), 2.9 mEq/L.

Seven months after diagnosis, he was referred to our hospital at which time he was examined by physical therapy. His voice sounded nasal. Swallowing was disturbed for solid materials. The patient demonstrated muscular fasciculation, atrophy, and weakness in the tongue. The Gag reflex was decreased, in particular on the left side. Position of uvula was deviated to the right side (curtain sigh). Laryngeal elevation was delayed during deglutition. He could benefit from head rotation to the left side to prevent aspiration. Head rotation can divert material down the opposite pyriform sinus into the esophagus.

Videofluoroscopic study showed (1) delayed bolus propulsion in the oral phase, (2) residue in velleculae after swallow, and (3) removal of the residue by repeated swallowing.

Pulmonary examinations revealed 3,500 mL of forced vital capacity (FVC) and 350 L/min of PCF. The patient was able to clear all tracheal secretions independently. Thus, cough function could be classified “functional”. Respiratory rate was 20 breaths per minute.

Sixteen months after diagnosis, he agreed to undergo tracheostomy. Nineteen months after diagnosis, he required mechanical ventilation.

METHODS

The author investigated longitudinal changes of the peak expiratory flow by two different methods and FVC. The two different methods were coughing and huffing (FET).

Peak expiratory flow was measured with a peak flow meter (Assess; Health scan products Inc; Cedar Grove, NJ). FVC was measured with the Wright Spirometer (Mark 14, Ferraris Development and Engineering Co, Ltd, London UK). A facemask was connected to the measurement devices (Fig. 1). Peak expiratory flow and FVC were measured three times per session and the maximum value of three measurements was taken. Measurements were performed in the sitting position to eliminate the effect of postural dependent upper airway obstruction due to bulbar muscle involvement.
RESULTS

Decrease in PCF progressed over time (Fig. 2). Approximately 12 months after diagnosis, PCF decreased to 140 L/min, and the patient could not cough out the airway secretions independently. However, at this time, FET generated 250 L/min of peak expiratory flow. The peak expiratory flow was consistently greater in FET compared to coughing (Fig. 2). Decrease in PCF persisted, but FET maintained peak expiratory flow above 160 L/min until 14.5 months after diagnosis.

However, 15 months after diagnosis, FET generated a peak expiratory flow only 140 L/min. At this time, FVC had decreased to 1,600 mL (Fig. 3).

DISCUSSION

Cough flows less than 160 L/min are ineffective\(^{15}\). Approximately 12 months after diagnosis, PCF could not exceed 160 L/min. As a result, cough became ineffective.

The results of this case study revealed that peak expiratory flow was improved by FET, even though the patient with ALS demonstrated marked PCF decrease due to bulbar involvement.

Despite having bulbar involvement, the patient who had adequate both inspiratory and expiratory muscles could generate peak expiratory flow to a point over 160 L/min threshold until 14.5 months...
after diagnosis. As a result, FET delayed the need for tracheostomy.

Thus, FET is beneficial for patients with ALS and possibly other neuromuscular disorders which weaken the bulbar muscle. Nonetheless, FET maintained peak expiratory flow for a prolonged period of time until marked weakness from the progressive effects of ALS produced an inadequate contraction of the ventilatory muscles and a subsequent deterioration in FVC. FET, therefore, may be an important adjunctive treatment for patients with ALS and other neuromuscular disorders who have been diagnosed with bulbar involvement.

However, when the FVC was observed to be markedly decrease, and it was only 1,600 mL, the patient was unable to reach 160 L/min of peak expiratory flow by FET. The decrease of FVC was associated with a decrease in peak expiratory flow generated by FET, because peak expiratory flows can decrease from not only bulbar dysfunction but also inspiratory or expiratory muscles weakness. When the peak expiratory flow generated by FET declined to below 160 L/min, the patient required tracheostomy.

In addition, when FET became ineffective, weakness of deltoids, biceps and neck extensors became apparent, and the arterial blood gas values indicated alveolar hypoventilation.

The lower motor neuron cell bodies for the diaphragm are in the upper portion of the cervical spinal cord, segments C3 through C5. Lower motor neurons to neck extensors, deltoids, and biceps originate from the same cervical region. Thus, these muscles usually become weak with the diaphragm. Therefore, weakness of deltoids, biceps, and lower cervical-upper thoracic paraspinal muscles predicts involvement of respiratory muscles.

The effect of FET was dependent on FVC. Thus, appearance of severe weakness less than poor of deltoids, fair of biceps, and fair of neck extensors may be good indication that the beneficial time for FET in the patients with bulbar onset ALS is over.

The rate of neuronal deterioration in patients with ALS appears to vary. Therefore, the most beneficial time in the stage of the disease for FET for patients with ALS may be different among individuals. This is the major limitation of this study. However, the author believe that, when cough becomes ineffective in patients with bulbar onset ALS, physical therapists should instruct FET to delay the need for tracheostomy, and because FET is a method which can be performed by the patient independently to eliminate bronchial secretions.

REFERENCES