Long-term effect of air stacking using a respirator in Duchenne-type muscular dystrophy

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Abstract

Convalescence and respiratory function testing of five Duchenne-type muscular dystrophy patients who had continued with air stacking using a respirator for eight years were examined. Although artificial respiration management was received throughout the day, all five patients maintained some activities of daily living. Four of these Duchenne-type muscular dystrophy patients who were not able to continue with air stacking using a respirator were investigated as a disease control. Two of these four disease control patients died, and the remaining two cases regularly needed to lie in bed. The maximal forced air intake was maintained at 900 ml - 980 ml in the test cases, but was just above half that for the disease control case. The lung capacity was reduced by half in the test cases, but could not be measured for the disease control case. Air stacking using a respirator continues for a long period, and our results show that this treatment is effective in terms of maintaining the activities of daily living and the lung capacity of Duchenne-type muscular dystrophy patients.

Key Words: Duchenne-type muscular dystrophy, Air stacking, Artificial respiration management, Respirator, Lung capacity

Introduction

Among patients with Duchenne muscular dystrophy (DMD) who die of ventilatory failure, the majority of deaths are due to breathing muscle weakness. However, this tendency was changed by noninvasive positive pressure ventilation (NPPV), which began in the 1990s and showed positive results. For example, it was confirmed that the average lifespan of patients with DMD was extended by approximately ten years when NPPV was performed. In addition, respiratory care has become more and more important. In particular, breathing rehabilitation has attracted much attention. Exercise capacity decreases with increased systemic muscle weakness due to DMD, and joint contracture progresses simultaneously. This contracture extends to the thorax including, but not limited to, the four extremities. With breathing muscle weakness, reduced thoracic cage compliance causes a reduction in the ability to perform respiratory tract clearance. As a result, repeated atelectasis may occur, and patient prognosis can be greatly affected. In general, the main goals of breathing rehabilitation in DMD are maintenance of the lungs and thoracic cage compliance. When the subjects are children, it is important to prevent

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spinal column thoracic deformity and to maintain coughing ability at the same time. We conducted hyperinflation training for working on before, and good results were obtained. We studied whether hyperinflation training of the working upon using a respirator was effective in improving the long-term prognosis of patients with DMD.

Subjects and Methods

Clinical course was examined retrospectively in five patients with DMD who started treatment by air stacking using a respirator in 2000. In all cases, NPPV had already been performed. The age of the patients at that time ranged from 19 to 27 years old (average of 22.8+-3.2 years old). The air stacking involved a volume-cycled respirator, and ventilatory volume was gradually increased for a few days with a final aim of approximately 1,000 ml. The patients performed air stacking themselves with the final set ventilatory volume for around five minutes every morning thereafter. The interface involved a mouthpiece. A chief physician watched the attendant training for two weeks from the training start. The subsequent continuation of the procedure was left up to patients. When training was continued the staff only conducted setting changes and the training reached by oneself.

With regard to lung function measurement, the patients adopted a prostrate position on a bed, and, with a ventilation meter (Haloscale, Wright, UK) and a face mask, lung capacity (vital capacity, VC) and maximal forced air intake (maximum insufflation capacity, MIC) were measured. For patients that had received tracheostomy throughout their course, the measurement of respiratory function with a face mask was difficult. In these cases, respiratory function was measured by incorporating a ventilation meter in the respiratory circuit. The outcome of all cases was described, and VC and the MIC eight years later were compared with those at the training start time.

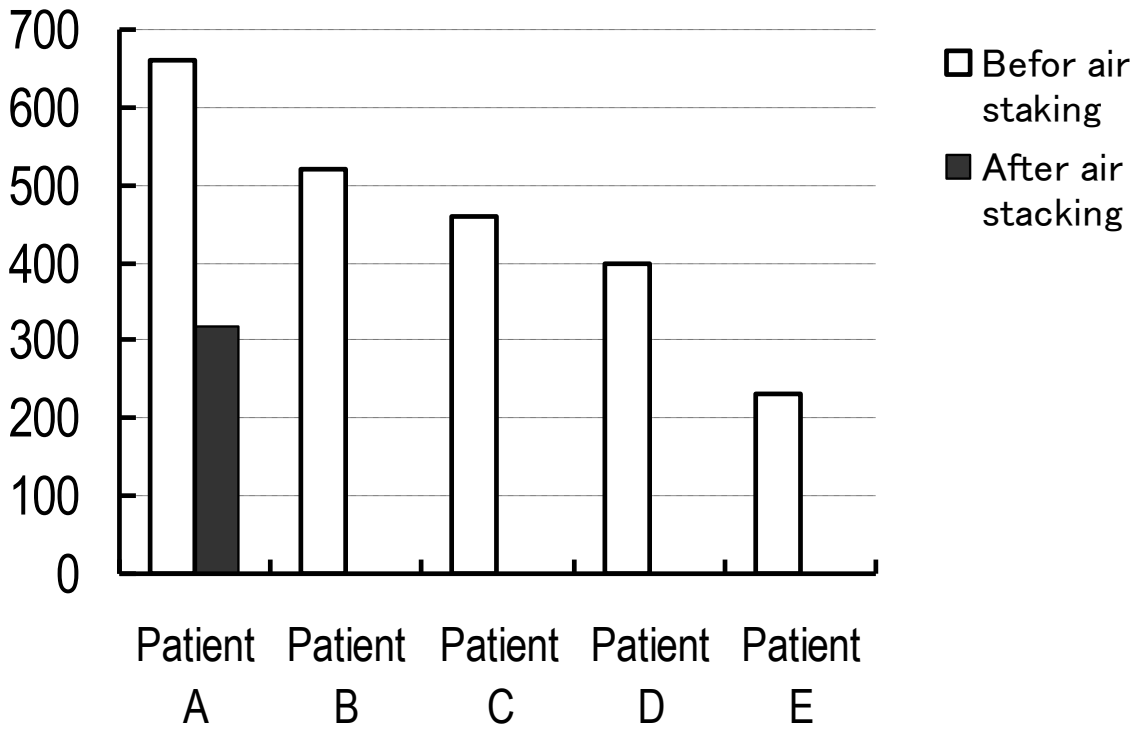

Figure 1. Changes in vital capacity before and after the air stacking manoeuvre. Only one of five patients (patient A) continued the training for eight years. Patients D and E died during the study.

Results

Changes in vital capacity (VC) before and after the air stacking manoeuvre are shown in Figure 1. Only one of the five patients (patient A) continued the training for eight years. The patient needed artificial respiration throughout the day but continued to carry out activities with an electric wheelchair during the days when NPPV was received. The four other patients chose to cancel the training within 1-2 months. Two patients who chose to cancel the training died seven (patient D) and eight years later (patient E). The four patients who chose to cancel the training all had aspiration pneumonia. The two surviving cases regularly lay in bed. Changes in MIC are shown in Figure 2. Because spontaneous respiration was extremely diminished in patients B and C with regard to VC, measurement with a face mask became impossible. Patients D and E died seven to eight years after the final assessment was performed.

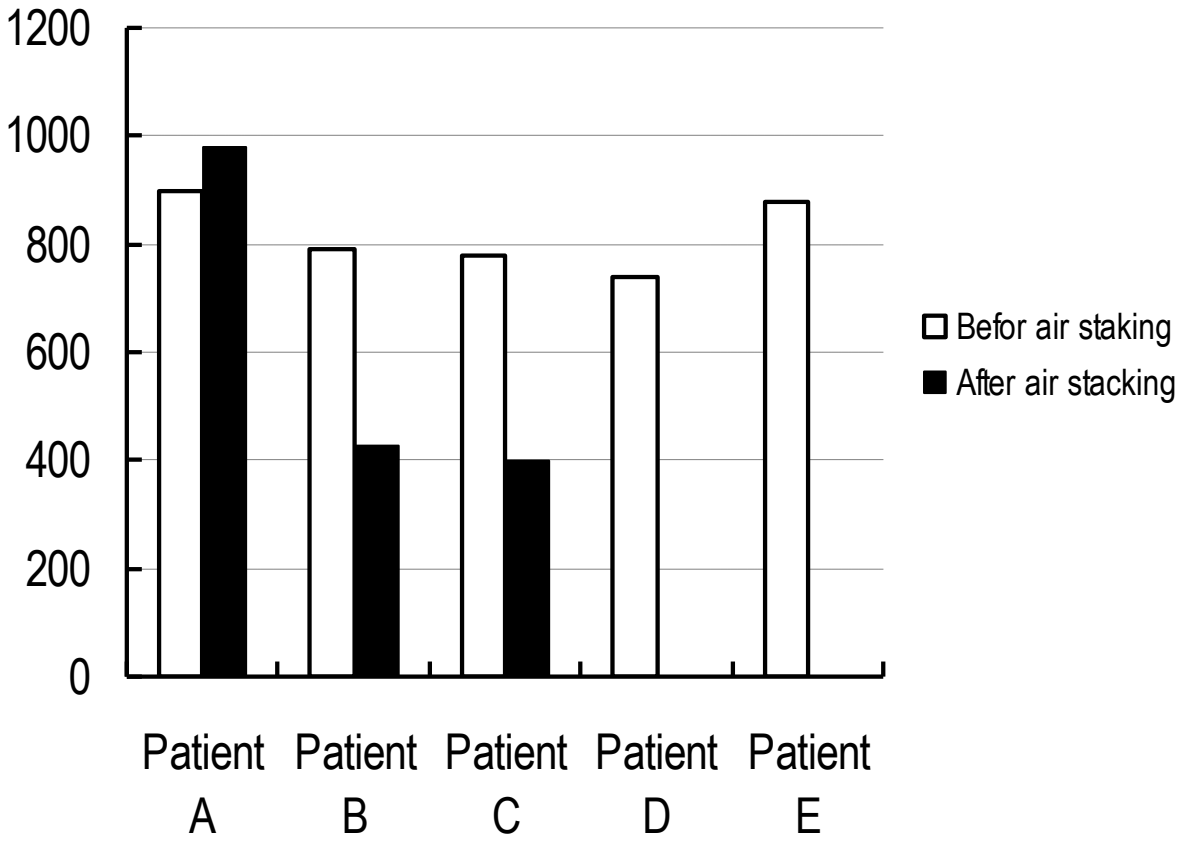

Figure 2. Changes in maximum insufflation capacity, MIC, were measured before and after the air stacking manoeuvre. MIC only increased in patient A. Patients D and E died during the study.

Discussion

The importance of respiratory tract clearance in the respiratory care of DMD patients has already been clearly described in management guidelines [1,2]. The most important goal of breathing rehabilitation in muscular dystrophy is respiratory tract clearance. Restrictive ventilatory defects are caused by a reduction in breathing muscular strength in neuromuscular disorders such as DMD, and lung capacity decreases. DeTroyer et al. [3] observed that lung capacity fell to less than the predicted amount in patients with neuromuscular disorders. They estimated that this difference was based on lung / thoracic cage compliance reduction. A physical therapist is involved from an early stage in preventing this process, and it is important to attempt compliance maintenance. Bach has described forced air-breathing methods using a volume-cycled respirator [4]. Our report showed the effectiveness of this method. Bach reported that it was important for the discharge of sputum to

ensure that there was an MIC of more than 1,000 ml. [5]. We therefore aimed to achieve an MIC of 1,000 ml, and the effectiveness of this approach was confirmed. For a progressive disease such as DMD, we cannot do anything for VC decreasing with a year. On the other hand, MIC can be maintained or increased by devising appropriate training. Our experience shows the effectiveness of these methods, and they should be examined more widely in the future.

References

1. Finder JD, Birnkrant D, Carl J, et al: Respiratory care of the patient with Duchenne muscular dystrophy: ATS consensus statement. Am J Respir Crit Care Med. 2004;15: 456-65,.

2. Kravitz RM. Airway clearance in Duchenne muscular dystrophy. Pediatrics 2009; 123: S231-S235.

3. De Troyer A, Borenstein S, Cordier R.Analysis of lung volume restriction in patients with respiratory muscle weakness. Thorax. 1980;35:603-10.

4. Bach JR. Respiratory considerations. In Guide to the evaluation and management of neuromuscular disease. Philadelphia: Handley & Belfus, 1999; 67-87

5. Bach JR, Alba AS. Management of chronic alveolar hypoventilation by nasal ventilation. Chest. 1990 Jan;97(1):52-7