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Review of respiratory muscle training in neuromuscular diseases

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In many neuromuscular diseases e.g. Duchene muscular dystrophy (DMD) and Spinal muscular atrophy (SMA), respiratory complication is one of the most commonly associated conditions as the diseases progress. The progressive weakness of the respiratory muscle could result in deterioration of ventilatory function of the respiratory system.[1] Respiratory muscle training (RMT) has been widely employed as an important component in the physiotherapy respiratory care for these groups of patients. The treatment aims to improve respiratory function by means of exercising the respiratory muscles. Nevertheless, controversial results were found in previous studies regarding the effectiveness of RMT to improve the respiratory muscle strength and/or endurance.[2-4] Direct comparison between studies is difficult because outcome measures and training protocol are not standardised. However, there are implications which are useful in our clinical practice.

Parameters in RMT evaluation

As many investigators studied the effectiveness of RMT by measuring the respiratory muscle strength and endurance, appropriate outcome measures should be chosen for evaluation purpose. The most commonly used measurement in determining the respiratory muscle strength is the maximal inspiratory pressure (MIP).[2,3,5,6] It is the maximum pressure that is produced when the subject is asked to make a maximal inspiratory effort against a completely occluded airway. For respiratory muscle endurance, the 12 seconds maximum voluntary ventilation (MVV) is measured.[3,4,6] It is the maximum ventilation that the subject can produce in 12 seconds. Apart from research purpose, the above mentioned two measurements are used very often in clinical practice. They are simple in administration, and involved easily understandable procedures even in young children.

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In the paediatric unit that the author is working in, MIP and MVV are measured as routine monitoring of respiratory muscle function in patients with neuromuscular disease.

RMT protocol

To design an effective training protocol is a challenge to clinicians when conducting a RMT programme. It aims to train respiratory muscles by carefully induced fatigue. The protocol should be simple and concise, yet not too boring in order to improve compliance. This involves a balance among several factors like number of repetitions, the amount of resistive loading during the breathing exercise, and the frequency of training sessions. For the training of respiratory muscle endurance, Koessler suggested a programme consisting 10 inspiratory resisted breathing cycles, each cycle lasting about 1 minute with 20 seconds rest between cycles.[3] It means that each training session will last around 15 minutes. Previous studies suggested the training programme should be performed twice daily.[3,5] However, the inspiratory resistance in these studies varied from 30%[5] to 70%[3] of the pre-determined MIP, with the resistance being adjusted accordingly as the MIP was reviewed every 3 months.[3] For training respiratory muscle strength in Koessler's study, the inspiratory resistance was set as high as 90% of the MIP, with only 10 breaths in each training session.[3] However, the problem of fatigue in this kind of training involving submaximal muscle work is questionable. The author adopts a low inspiratory resistance protocol for the endurance training with MIP starting at 30-40% of the pre-determined value. In clinical practice, it is a long term training programme with regular review of the respiratory function of the patients.

Patient selection for RMT

Apart from the structure of the training protocol, the effectiveness may be affected by the degree of decline in respiratory function of patients with neuromuscular

diseases. It has been shown that RMT effects was positively correlated with training intensity in patients with less than 10% decline in respiratory function during the year before the training began.[6] In patients whose respiratory function decline exceeded 10% in the year prior to training, the effect was independent of the training intensity. In another study, in patients with vital capacity of less than 25% predicted and/or $PaCO_2$ values of more than 45 mmHg, there was no improvement in respiratory function after 1 month of training.[7] For patients with better respiratory function parameters, significant improvement was found. However, the number of subjects was small in the two above-mentioned studies; and their results should be interpreted with caution. A reasonable assumption is that it is more beneficial to perform RMT in patients with lesser degrees of decline in respiratory function, hence early intervention is recommended.

In summary, RMT protocol involves inspiratory loaded breathing exercises. The optimum resistance varies according to different studies. The use of high resistance in training respiratory muscle strength is controversial. Moreover, patients with different levels of respiratory function may show different response to the training. A training programme tailor-made for the individual with regular review is suggested for patients with neuromuscular diseases as their underlying condition worsens. Further studies in the variables of the protocols are warranted in order to improve their effectiveness.

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