Cochrane Corner

Does respiratory muscle training improve respiratory function compared to sham training, no training, standard treatment or breathing exercises in children and adults with neuromuscular disease? A Cochrane Review summary with commentary

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Abstract.
BACKGROUND: Progressive muscle weakness is a feature of neuromuscular diseases (NMDs), a heterogeneous group of conditions with variable onset, presentation and prognosis that affect both children and adults. Respiratory muscle weakness compromises respiratory function and may lead to respiratory failure.
OBJECTIVE: To assess the effects of respiratory muscle training (RMT) in adults and children with NMD.
METHODS: A Cochrane Review by Silva et al. was summarized with comments.
RESULTS: Eleven studies involving 250 randomized participants with NMD were included. While the studies showed that RMT may lead to improvements in lung function and respiratory muscle strength in people with ALS and DMD, this was not a consistent finding. The evidence from all the included trials was of low or very low certainty.
CONCLUSIONS: There may be some improvement in lung capacity and respiratory muscle strength following RMT in some NMD. There appears to be no clinically meaningful effect of RMT on physical functioning and quality of life in ALS. The low certainty of the evidence means that the results need to be interpreted with caution.

Keywords: Neuromuscular disease, respiratory muscle weakness
The aim of this commentary is to discuss in a rehabilitation perspective the published Cochrane Review “Respiratory muscle training in children and adults with neuromuscular disease” by Silva et al.1, under the direct supervision of the Cochrane Neuromuscular Group. This Cochrane Corner is produced in agreement with Neurorehabilitation by Cochrane Rehabilitation.

1. Background

Neuromuscular diseases (NMD) are a broad group of disorders that may affect any level of the neuromuscular system, including muscle, neuromuscular junction, upper and lower motor neurons or any combination of these (Morrison et al., 2016). They may be hereditary (usually affecting children) or acquired (mainly in adults), and slowly or rapidly progressing, with a prevalence of the majority of NMD of 1–10 per 100,000 population (Deenen et al., 2015). These diseases are characterized by progressive muscle weakness, with respiratory muscle weakness as a major cause of morbidity and mortality, manifesting as impaired ventilatory function as a result of compromised breathing, airway patency or cough (Voulgaris et al., 2019). Respiratory muscle training (RMT) may improve strength and endurance of inspiratory or expiratory muscles and may improve respiratory function (Pfeffer & Pfitz, 2016).

Respiratory muscle training in children and adults with neuromuscular disease
(Silva et al., 2019)

1.1. What were the aims of this Cochrane Review?

The aim of this Cochrane Review was to assess the effects of respiratory muscle training (RMT) in adults and children with neuromuscular disease (NMD).

1.2. What was studied in the Cochrane Review?

The population addressed in this review included adults (>18 years) and children of both sexes with a diagnosis of neuromuscular disease (NMD) of any degree of severity. Participants were community-dwelling and not requiring mechanical ventilation, and included those with myopathies, disorders of the neuromuscular junction and neuropathies. Those with acute respiratory failure and cognitive impairment were excluded. The intervention studied was RMT compared to sham training, no training, standard treatment, breathing exercises or other intensities or types of RMT. The primary outcomes were measures of lung capacity; secondary outcomes were inspiratory and expiratory muscle strength, physical function, quality of life, unscheduled hospitalizations for chest infection or acute exacerbations of chronic respiratory disease, and adverse events.

1.3. Search methodology and up-to-dateness of the Cochrane Review?

The review authors searched the Cochrane Neuromuscular Specialised Register, the Cochrane Central Register of Controlled Trials. MEDLINE, Embase on 19 November 2018, and the US National Institutes of Health Clinical Trials Registry and the World Health Organization International Clinical Trials Registry Platform on 23 December 2018 to identify RCTs and quasi-RCTs, including crossover trials of RMT. They also searched reference lists and relevant manufacturers’ websites for trial information.

2. What are the main results of the Cochrane Review?

Eleven studies involving 250 randomized participants with NMD were included, six of which involved participants with Duchenne Muscular Dystrophy (DMD, n = 112), three involving people with Amyotrophic Lateral Sclerosis (ALS, n = 88), and one trial each in people with Becker or limb girdle muscular dystrophy and myasthenia gravis (n = 23 and n = 27 respectively). Thirteen participants were excluded from the analysis by the trialists. No trials provided long-term data (>1 year). Eight studies involved inspiratory muscle training, four using training loads between 15% to 60% of maximal inspiratory pressure, and four trials using resistive training. Two studies involved expiratory muscle training, and one
trial used a combined strength and endurance training protocol. Nine trials were considered at high risk of bias in at least one of the seven domains, and different outcome measures used precluded meta-analysis. Only one study reported on adverse events. While the studies showed that RMT may lead to improvements in lung function and respiratory muscle strength in people with ALS and DMD, this was not a consistent finding. In people with ALS, two studies examined the effect on physical function and one trial assessed quality of life and indicated that there was no clear effect. The evidence from all the included trials was of low or very low certainty.

2.1. What did the authors conclude?

The authors concluded that lung capacity and respiratory muscle strength may be improved following RMT in ALS and DMD although this was inconsistent. There appears to be no clinically meaningful effect of RMT on physical functioning and quality of life in ALS. The low certainty of the evidence means that the results need to be interpreted with caution.

2.2. What are the implications of the Cochrane evidence for practice in neurorehabilitation?

The level of evidence is insufficient to determine whether RMT is effective in improving respiratory function compared to sham training, no training, or breathing exercises. If RMT were to be prescribed for individual patients with NMD, the effects on lung function and any adverse events would need to be carefully monitored. Development of clinical practice guidelines on RMT depends on further well-conducted research. It is highly desirable that clinicians and researchers fully record all components of interventions, e.g. using the TIDieR checklist (Hoffmann et al., 2014), to improve reporting, and for future implementation in the clinical setting. Assessment of physical function (activity and participation) is an important outcome in rehabilitation in addition to measures of respiratory function (impairment), and careful follow-up of adverse events.

References


