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Randomized trial of lung hyperinflation therapy in children with congenital muscular dystrophy

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Abstract

Objective: Respiratory compromise in congenital muscular dystrophy (CMD) occurs, in part, from chest wall contractures. Passive stretch with hyperinsufflation therapy could reduce related costo-vertebral joint contractures. We sought to examine the impact of hyperinsufflation use on lung function and quality of life in children with CMD.

Study design: We conducted a randomized controlled trial on hyperinsufflation therapy in children with CMD at two centers. An individualized hyperinsufflation regimen of 15 minutes twice daily using a cough assist device over a 12 months period was prescribed. We measured lung function, quality of life, and adherence. To demonstrate reproducibility, pulmonary function was measured twice on the same day. A mixed-effects regression model adjusting for confounders was used to assess the effects of hyperinsufflation.

Results: We enrolled 34 participants in the study; 31 completed the trial (n = 17 treatment group and n = 14 controls). Participants in the treatment group demonstrated a relative gain in lung volume measured at 4 and 8 months, but not at 12 months. The control group required increases in the maximum insufflation pressures to achieve maximum lung volumes while the treatment group did not. Adherence was best early in the study, peaking at the first visit and decreasing at subsequent visits. Caregiver-reported quality of life was higher in the treatment group.

Conclusion: Hyperinsufflation therapy is effective in increasing and sustaining lung volume over time. Adherence, however, was inconsistent and difficult to maintain. Further research should determine if improved adherence leads to sustained benefits of hyperinsufflation.

Keywords: adherence; neuromuscular disease; respiratory insufficiency.

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