



Respiratory Implications of Pediatric Neuromuscular Disease.

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ABSTRACT :

Children with progressive neuromuscular weakness undergo a stereotypical progression of respiratory involvement, beginning with impaired airway clearance and progressing to nocturnal and then diurnal ventilatory failure. This review examines issues related to airway clearance and mucus mobilization, sleep problems, and use of assisted ventilation in children with neuromuscular diseases. Interventions for each of these problems have been created or adapted for the pediatric population. The use of airway clearance therapies and assisted ventilation have improved survival of children with neuromuscular weakness. Questions regarding the best time to introduce some therapies, the therapeutic utility of certain interventions, and the cost-effectiveness of various treatments demand further investigation. Studies that assess the potential to improve quality of life and reduce hospitalizations and frequency of lower-respiratory tract infections will help clinicians to decide which techniques are best suited for use in children. As children with neuromuscular disease survive longer, coordinated programs for transitioning these patients to adult care must be developed to enhance their quality of life.

KEYWORDS:

airway clearance therapies; high-frequency chest-wall compressions; intrapulmonary percussive ventilation; mechanical in-exsufflator; noninvasive ventilation; sleep-disordered breathing

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