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Respiratory management using Cough Assist in a toddler with spinal muscular atrophy type 1: a case study

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Introduction

Spinal muscular atrophy type 1 (SMA-1) is a rare disease. The estimated incidence is 1 in 10,000 live births. Typical natural history is respiratory failure and death before 2-year old if without respiratory intervention. Respiratory muscle weakness results in impaired cough, inability to clear lung secretions, chest underdevelopment and hypoventilation. With new advances in respiratory management, quality of life (QoL) and survival time improve. Cough Assist is a non-invasive therapy using mechanical insufflation-exsufflation. Various clinical guidelines recommend neuromuscular patients with ineffective cough to use it. However, evidence to support its application in toddlers with SMA-1 is limited.

Objectives

This case study evaluates the impact on respiratory function and QoL in a toddler with SMA-1 after using Cough Assist therapy at the age of 25-month.

Methodology

This was a prospective pre-post case study. Assessments were performed at baseline and 5 months post-treatment. Outcome measures include frequency of chest physiotherapy and suction, chest deformities, Pediatric Quality of Life Inventory Neuromuscular Module 3.0 (PedsQL-NM), respiratory admission and length of stay (LOS).

Result

The subject was a boy who presented significant hypotonia and gross motor delay at 3-month old. Muscle biopsy confirmed the diagnosis of SMA-1 at 19-month old. Nocturnal BIPAP ventilation, home chest physiotherapy and suctioning started at 20-month old. Home-based Cough Assist machine began at 25-month old, adapting the protocol of the University of Wisconsin with the inhale and exhale pressures titrated gradually to 30 cmH₂O. Pre- and Post-treatment Comparisons: 1. The chest

shape progressed from “bell-shape” to “square-shape” with decreased pes excavatum deformity. 2. Daily duration of chest physiotherapy reduced 50%, from 2 hours to 1 hour. Daily frequency of suction reduced 50%, from 8 times to 4 times. 3. There were 3 respiratory admissions with 49 days of LOS in the previous 5-month pre-treatment. There was no respiratory admission post-treatment. 4. PedsQL-NM measures health-related QoL. The score increased from 30 to 34 (out of 100), indicating better QoL. The results demonstrated that Cough Assist therapy was effective in removing lung secretions, reducing respiratory infections and hospital admission in the 25-month old boy with SMA-1. The scores of PedsQL-NM indicated improvement in QoL, which was well echoed by the feedback of the family.