Airway Clearance Indications: Neuromuscular/Neuromotor Disorders

Spinal Muscular Atrophy (SMA)

Spinal muscular atrophy is a term applied to a number of genetic disorders affecting the motor neurons of the brain stem and spinal cord. Also called anterior horn cells, these neurons are a crucial communication pathway from the brain stem and spinal cord to the muscles. Muscles rely on input from the motor neurons for basic function. If that input is inadequate or inaccurate, muscle function is severely impaired and muscle wasting—atrophy—occurs. Bulbar (swallowing), respiratory and limb muscles are predominantly affected.

There are dozens of disorders affecting the motor neurons, but spinal muscular atrophy is distinguished by its genetic nature and early onset. There are at least three distinct categories of spinal muscular atrophy characterized by severity and age of first symptoms. Infantile (type I) spinal muscular atrophy, also called Werdnig-Hoffmann syndrome, is the most severe form of the disorder with onset between birth and six months and a life expectancy of less than one year. Intermediate, or type II, spinal muscular atrophy generally has an onset between ages 7 and 18 months. Children with this form of the disorder may never be able to stand or walk on their own, but they may have the strength to sit on their own at some point. Eventually, however, progressive muscle weakness will result in wheelchair reliance. Individuals with type III spinal muscular atrophy, also called Kugelberg-Welander disease, exhibit milder symptoms and can live into adulthood. Chronic respiratory complications are the primary cause of illness and death in all forms of spinal muscular atrophy.

The SMA Foundation estimates that there are 25,000 Americans with spinal muscular atrophy. It is the leading genetic cause of death in children.

What Happens in Spinal Muscular Atrophy?
Gradual destruction of both upper (found in the brain) and lower (found in the brain stem and spinal cord) motor neurons results in progressive weakness, spasticity and paralysis of the muscles, including those that control speech, swallowing, facial expressions, voluntary movement of the extremities and respiration As paralysis progresses, respiratory status is compromised and the majority of affected individuals succumb to death by respiratory failure within three to five years of diagnosis.

Ineffective cough due to weakened respiratory muscles and the inability to manage oral secretions due to inadequate bulbar (swallow) function can contribute to impaired airway clearance. Oral secretions that can't be swallowed may be aspirated into the airways overwhelming the mucociliary clearance system. Ineffective cough compounds the problem and a vicious cycle of secretion retention, infection, inflammation and airway damage may set in. Additionally, in advanced disease, individuals with amyotrophic lateral sclerosis require ventilatory support. Ventilator-associated pneumonia and other respiratory infections are well-documented complications of prolonged ventilator dependence and create additional risk factors for

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compromised airway health in amyotrophic lateral sclerosis.

How Airway Clearance Therapy Can Help Amyotrophic Lateral Sclerosis
There is currently no cure for amyotrophic lateral sclerosis so therapy is targeted at improving quality of life and daily function. Keeping the airways clear of excess secretions and thereby reduce the incidence of inflammation and/or infection and is crucial to maintaining respiratory health. Airway clearance therapy using High Frequency Chest Wall Oscillation (HFCWO) has been demonstrated by clinical study to promote excess mucus clearance and improve bronchial drainage. Shear forces are created by HFCWO treatment that mechanically releases adhered secretions from the walls of the pulmonary tract. HFCWO has also been shown to reduce the viscosity of secretions which significantly improves mobilization of excess mucus. By replicating cough, HFCWO can effectively mobilize pulmonary secretions from smaller airways to larger airways where they can be coughed out, swallowed or suctioned.

Symptoms of Amyotrophic Lateral Sclerosis

- Twitching and cramping of muscles, especially those in the hands and feet
- Weakness and/or loss of motor control in the hands and arms
- General weakness and fatigue
- Tripping and falling
- Dropping things
- Uncontrollable periods of laughing or crying
- Slurred or thick speech and difficulty in projecting the voice
- Shortness of breath
- Difficulty breathing
- Difficulty swallowing
- Paralysis

For More Information on Spinal Muscular Atrophy:

1. The SMA Foundation: http://www.smafoundation.org/

2. Fact Sheet on SMA from the National Institute of Neurological Disorders and Stroke (NINDS): http://www.ninds.nih.gov/disorders/sma/sma.htm

3. National Library of Medicine Fact Sheet on SMA: http://ghr.nlm.nih.gov/condition=spinalmuscularatrophy

4. Fact Sheet on SMA from The Children's Hospital of Philadelphia: http://www.chop.edu/consumer/your_child/condition_section_index.jsp?id=-9273

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