

## **Airway Clearance Indications: Neuromuscular/Neuromotor Disorders**

### **Spinal Muscular Atrophy (SMA)**

Spinal muscular atrophy (SMA) is a term applied to a number of genetic disorders affecting the specialized nerve cells, called motor neurons, of the part of the brain that is connected to the spinal cord, the brain stem, and spinal cord itself. 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. SMA is divided into subtypes, which are based on the age when symptoms appear and the severity of the disease.

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 SMA?**

There are dozens of disorders affecting the motor neurons, but SMA is distinguished by its genetic nature and early onset. There are at least three distinct categories of SMA characterized by severity and age of first symptoms. Infantile (type I) SMA, 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, SMA 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 SMA, 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.

#### **How Airway Clearance Therapy Can Help SMA**

There is currently no cure for SMA 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 SMA

- Muscle atrophy
- Loss of motor function
- Increasing weakness
- Reduced strength
- Limited function and mobility
- Bone and/or spinal deformities, which may require surgical treatment
- Respiratory illnesses, possibly severe
- Chronic respiratory complications

## 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](http://www.chop.edu/consumer/your_child/condition_section_index.jsp?id=-9273)