SmartVest® AIRWAY CLEARANCE SYSTEM

Airway Clearance Indications:

Muscular Dystrophy

Muscular dystrophy is a term used to describe a diverse group of inherited disorders primarily affecting the muscles. Age of onset, severity of disease and rate of progression are determined by the specific genetic abnormality and type of muscular dystrophy. Most muscular dystrophies are progressive, but the rate of disease progression differs based on the type of muscular dystrophy. Duchenne muscular dystrophy, the most common and most severe of the muscular dystrophies, is generally passed as an x-linked trait from mother to son. It occurs with a frequency of 1 in every 3,500 births. Children with Duchenne muscular dystrophy experience rapidly progressing disability and are usually wheelchair-dependent by age 10 to 12. Becker muscular dystrophy, the second most common muscular dystrophy, generally progresses more slowly and occurs with a frequency of 1 in 30,000 births.

What Happens in Muscular Dystrophy?
The underlying cause for the majority of muscular dystrophies is the absence or faulty production of a crucial protein called dystrophin which is required to build and maintain muscle mass and function. Muscular dystrophies related to genetic disorders of dystrophin are also known as dystrophinopathies. The absence of dystrophin results in a breakdown of muscle tissue and the inability to maintain the strength and stability of the muscle. Muscle tissue is replaced by fibrous (scar) tissue and fatty tissue and becomes weak and unstable. The muscular dystrophies may involve the heart muscles, breathing or respiratory muscles, and other body systems such as the gastrointestinal tract. Weak, unstable muscle tissue is unable to support the skeletal structure and gait (walking) abnormalities and serious spinal curvature (scoliosis) develop. Eventually, voluntary motion is severely limited and affected individuals become confined to a wheelchair, an event that greatly accelerates the rate of scoliosis.

Muscular dystrophy-related scoliosis in which the spine and thorax (chest cavity) are impaired by structural instability restrict the ability of the lungs to expand and contract fully, a condition known as restrictive lung disease. Restrictive lung disease creates a disruption in air flow, characterized by reduced lung volumes (total amount of air contained in the lungs during breathing) and vital capacities (ability of the lungs to effectively use that air). In diseases where respiratory muscles are weak and the spine and thorax deformed, the ability to take in sufficient air may be constrained by the physical deformity. Insufficient inhalation may impair the ability to generate the expiratory force necessary to create an effective cough, resulting in atelectasis (collapse of the small airways and/or alveoli) and mucus plugging. Gastroesophageal reflux may cause aspiration of gastric contents into the lungs, adding to airway clearance burden. In conditions where muscle tone is weakened cough clearance is further compromised and repeated respiratory infections and pneumonia are common. As a result, the pulmonary mucociliary clearance system may become overwhelmed, resulting in a vicious cycle of secretion retention, infection, inflammation and airway damage.

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Rev 3/27/2009

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How Airway Clearance Therapy Can Help Muscular Dystrophy
There is currently no cure for muscular dystrophy. Therapeutic goals focus on maximizing function and improving quality of life. 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 Muscular Dystrophy:

- Numbness or weakness in one or more limbs, which typically occurs on one side of your body at a time or the bottom half of your body
- Partial or complete loss of vision, usually in one eye at a time, often with pain during eye movement (optic neuritis)
- Double vision or blurring of vision
- Tingling or pain in parts of your body
- Electric-shock sensations that occur with certain head movements
- Tremor, lack of coordination or unsteady gait
- Fatigue
- Dizziness

For More Information on Muscular Dystrophy:

1. Muscular Dystrophy Association: http://www.mda.org/

2. Fact sheet from the National Institute of Neurological Disorders and Stroke (NINDS): hhttp://www.ninds.nih.gov/disorders/md/md.htm

3. Fact sheet from the Mayo Clinic: http://www.mayoclinic.com/health/muscular-dystrophy/DS00200

4. Fact sheet from the American Academy of Orthopedic Surgeons: http://orthoinfo.aaos.org/topic.cfm?topic=a00384

5. Fact sheet from Cleveland Clinic: http://my.clevelandclinic.org/disorders/Muscular_Dystrophy/hic_Muscular_Dystrophy.aspx

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