Amyotrophic lateral sclerosis (ALS, also known as Lou Gehrig’s Disease or motor neuron disease) is a progressive neurodegenerative disease that affects nerve cells in the brain and the spinal cord. Motor neurons reach from the brain to the spinal cord and from the spinal cord to the muscles throughout the body. When the motor neurons die, the ability of the brain to initiate and control muscle movement is lost. With voluntary muscle action progressively affected, patients in the later stages of the disease may become totally paralyzed. The progressive degeneration of the motor neurons in ALS eventually leads to death. There is currently no cure for ALS.

Based on U.S. population studies, a little over 5,600 people in the U.S. are diagnosed with ALS each year. It is estimated as many as 30,000 Americans have the disease at any given time. According to the ALS CARE Database, 60% of the people with ALS in the database are men and 93% of patients in the database are Caucasian.

Most people who develop ALS are between the ages of 40 and 70, with an average age of 55 at the time of diagnosis. Generally, ALS occurs in greater percentages as people grow older. Half of all people affected with ALS live at least three or more years after diagnosis. Twenty percent live five years or more; up to ten percent will live more than ten years.

Forms:

The most common form of ALS in the United States is "sporadic" ALS, 90 to 95% of all cases. It may affect anyone, anywhere. "Familial" ALS (FALS) means the disease is inherited or occurring more than once in a family lineage (genetic dominant inheritance). FALS accounts for a very small number of cases in the United States - 5 to 10% of all cases. In FALS, there is a 50% chance each offspring will inherit the gene mutation and may develop the disease.

ALS & NeuRx®:

One of the most important effects of progressive neuromuscular weakness in patients with ALS is the effect of the disease on respiration. Although ALS has no direct effect on the lungs, it has devastating effects on the mechanical function of the respiratory system. ALS affects all of the major respiratory muscle groups: upper airway muscles, expiratory muscles, and inspiratory muscles. Therefore, all patients with ALS are at significant risk for respiratory complications. Progressive inspiratory muscle weakness in ALS inevitably leads to carbon dioxide retention, inability to clear secretions and hypercarbic respiratory failure, which is the major cause of death in ALS. Pulmonary complications and respiratory failure are reported to be responsible for 77% - 84% of deaths in ALS.
Clinical trials for the use of the NeuRx Diaphragm Pacing System (DPS)® on patients ALS began in 2005. In the US, Synapse Biomedical is conducting a Post Market Surveillance study at a few select sites. The intent of the study is to expand on the safety and benefit body of knowledge. This includes the relationships with survival time, onset type (bulbar and limb), time from onset to treatment, and use with NIV, riluzole, or PEG when treated with the NeuRx DPS®.

In the United Kingdom, Synapse Biomedical is supporting a 108 person two arm clinical trial www.controlled-trials.com/ISRCTN53817913 to determine efficacy in prolonging life and maintains quality of life using the NeuRx DPS® in addition to current standard care non invasive ventilation (NIV).