Spinal Muscular Atrophy

About the Disorder: Spinal Muscular Atrophy (SMA) is a rare, inherited disease that is characterized by a progressive loss of muscle control, movement, and increased weakness due to the loss of motor neurons in the spinal cord. Proximal muscles (those c losest to the spine) are most severely affected. Cognitive ability, emotional development, and sensory nerves are unaffected. SMA has a wide range of severity but all people with SMA will either never acquire or will progressively lose the ability to walk, stand, sit, and eventually move. The age of onset of SMA varies but most individuals show symptoms of the disease during infancy or toddler years. Respiratory illness is more common in individuals with SMA, as are spinal and bone deformities. A quality multidisciplinary approach to care which can include physical therapy, occupational therapy, respiratory therapy, nutritional care etc. can improve quality and length of life for the individual with SMA.

Different Forms of SMA:

Type I; Acute SMA (Werdnig-Hoffman Disease): The general age on onset of Type I is between birth and 6 months. It is characterized by generalized muscle weakness, weak cry, trouble swallowing and suckling, breathing distress, and inability to sit without support. Type 1 generally progresses more quickly than the other types of SMA.

Type II: The generally age of onset is between 6 and 18 months. Muscles closest to the center of the body such as the shoulders, hips, thighs, and upper back are most severely affected. Respiratory muscles can also be affected and spinal curvature issues need to be monitored and treated appropriately. Type II usually progresses slowly.

Type III (Kugelberg-Welander Disease): The general age of onset is after 18 months and like the other forms muscles closest to the spine are most severely affected. The disease progresses slowly and the ability to walk can be maintained into adulthood and life span is generally not affected.

Most doctors consider the different SMA types to be on a continuum of severity and do not make rigid predictions about muscle weakness and life expectancy.