What is the adult form of Spinal Muscular Atrophy (SMA)

SMA is a motor neuron disease which typically presents in infancy and childhood with a known genetic mutation in the majority of cases. Progressive muscle weakness is caused by degeneration of lower motor neurons in the spinal cord and brainstem. The SMA phenotype ranges from a life threatening disease in the severely disabled infant to a mildly affected individual with a normal life expectancy. The mildest form of SMA in adulthood presents with weakness of the proximal musculature and functional impairments. This fact sheet will address adults living with SMA. (See Pediatric PT Section for SMA information in Infants and Children)

Motor symptoms:
- Hypotonia (lower than normal muscle tone)
- Proximal muscle weakness particularly the hip and shoulder musculature
- Impaired function including inability to run, altered gait pattern, difficulty rising from low surfaces (Gower’s maneuver) and climbing stairs
- Inability to raise arms over head or carry heavy objects
- In more severe forms, people with SMA can be wheelchair bound with only minimal hand function

Other symptoms frequently associated with the disease include:
- Skeletal deformities (scoliosis, hip dislocation, and joint contractures)
- Respiratory insufficiency
- Difficulty swallowing
- Fatigue
- Hand tremors
- Tongue fasciculations (muscle twitches)

Unique issues for adults with SMA are:
- Genetic counseling and family planning
- Equipment and self-care needs
- Assistive technology and accessibility
- Travelling with durable medical equipment
- Energy conservation and fatigue

There are no curative treatments available for SMA to date. In the absence of pharmacological therapies, clinical management is focused on prevention and treatment of complications due to muscle weakness. A multidisciplinary team of providers are needed to address genetic, pulmonary, nutritional, musculoskeletal, and rehabilitation issues.
How Can Physical Therapy (PT) Help adults with SMA?

The overall goal of physical therapy is to maximize function, strength and endurance through:

- Range of motion, positioning, and bracing to prevent contractures
- Moderate intensity exercise program which includes strengthening exercises in combination with aerobic exercise to maximize function and prevent deconditioning
- Mobility aids such as walkers and wheelchairs to maintain independence
- Aquatic or body weight supported therapy to promote movement not possible against gravity
- Energy conservation techniques
- Fall prevention programs
- Yoga and hippotherapy to augment standard therapeutic interventions as they promote flexibility and strength

References


Links to resources

http://www.fsma.org/FSMACommunity/UnderstandingSMA/
http://www.fightsma.org/
http://www.smafoundation.org/about-sma/