

## **Adventist Healthcare: Health Tip of the Week**

### **Childhood Onset SMA**

Spinal muscular atrophy (SMA) is the No. 1 genetic killer of children under the age of two. One in every 6,000 babies is born with SMA.

#### Description

If two unsuspecting carriers of the abnormal gene for SMA have children, the chance of a child inheriting the abnormal gene from each parent and thus developing the disease, is one in four.

\* The childhood SMAs are all autosomal recessive diseases; they run in families and more than one case is likely to occur in siblings or cousins of the same generation. Parents usually have no symptoms but still carry the gene.

#### Symptoms

SMA is a disease caused by progressive degeneration of motor neurons in the spinal cord.

\* The disorder causes weakness and wasting voluntary muscles. This affects crawling, walking, head/neck control and swallowing.

#### Three common types of SMA in childhood

\* Type I (Severe) Werdnig-Hoffman Disease: Diagnosis is usually made before six months of age. Child is unable to roll or sit unsupported. Also, has trouble breathing, sucking and swallowing, is weak and lacks motor development. Child usually does not live beyond 18 months of age.

\* Type II (Chronic): Diagnosis is made before two years of age. Affects infants between 7-18 months old. Child may be able to sit unaided or even stand with support. He/she usually does not suffer from feeding/swallowing difficulties but is at risk for respiratory infections.

\* Type III (Mild) Kugelberg-Welander Disease: Diagnosis is usually made after two years of age. It is the least deadly form of childhood-onset SMA. Child is able to walk, although with difficulty. He/she is able to stand but weakness is prevalent, eventually leading to a wheelchair.

#### Treatment

At this time, there is no cure for any types of SMA but researchers have found the specific gene that, when mutated, causes the disease.

\* Mildly-affected children may live into adult years. The more severely-affected children may die due to pneumonia and other chest problems before or in their teens.

\* Treatment is symptomatic and supportive. It includes treating pneumonia, curvature of the spine and respiratory infections. Physical therapy, ortho supports and rehabilitation are useful. Genetic counseling is imperative.

Sources: MDA, National Institute of Neurological Disorders and Stroke (NINDS), Families of SMA (FSMA) and Washington and Shady Grove Adventist Hospitals. The Health Tip of the Week is for educational purposes only. For additional information, consult your physician. Please feel free to copy and distribute this health resource.