

"Roussy-Lévy Syndrome, also known as hereditary areflexic dystasia, is a rare genetic neuromuscular disorder that typically becomes apparent during early childhood. The disorder is characterized by incoordination, poor judgment of movements (sensory ataxia), and absence of reflexes (areflexia) of the lower legs and, eventually, the hands; weakness and degeneration (atrophy) of muscles of the lower legs; abnormally high arches of the feet with increased extension of the toes (pes cavus or "clawfoot"); and tremors of the hands. Many affected individuals also have an abnormal front-to-back and sideways curvature of the spine (kyphoscoliosis). In individuals with Roussy-Lévy Syndrome, there is a failed communication of certain nerve signals to muscles of the lower legs (denervation). Roussy-Lévy Syndrome is inherited as an autosomal dominant genetic trait. Symptoms of Roussy-Lévy Syndrome are similar to other hereditary motor sensory neuropathies in that there is weakness and atrophy of the leg muscles with some loss of feeling. People with this syndrome have difficulty walking and a lack of reflexes and deformity of the foot or feet (pes cavus). Roussy-Lévy differs, however, from other hereditary motor sensory neuropathies because of the very early onset of the disorder during childhood and its slowly progressive course. Roussy-Lévy also has as one of its characteristics a slight tremor in the hands. Roussy-Lévy is a rare disorder that affects both sexes in equal numbers. Onset is during early childhood. Treatment of Roussy-Lévy Syndrome may include use of braces for the foot deformity or orthopedic surgery on the feet to correct the imbalance of the affected muscles. Genetic counseling may be of benefit to patients and their families. Other treatment is symptomatic and supportive."