Batten Turner Type Congenital Myopathy is an extremely rare, inherited muscle disease (myopathy) and is characterized by the lack of muscle tone or floppiness at birth (congenital hypotonia). The symptoms of Batten Turner Type Congenital Myopathy are slowly progressive during infancy and childhood. However, this disorder is not progressive in adulthood. The first symptoms of Batten Turner Type Congenital Myopathy in a newborn are the slow, progressive loss of muscle tone characterized by floppiness (hypotonia) and general weakness. Early motor skills and other important developmental milestones may be slightly delayed. Toddlers with this disorder usually have mild muscle weakness and may be prone to falling or stumbling. The muscles of the pelvis, neck, and shoulder area are most commonly affected. Since the symptoms of this disease are not progressive during adulthood, most people with Batten Turner Type Congenital Myopathy walk normally as adults. However, some physical activities may be slightly impaired. Batten Turner Type Congenital Myopathy is inherited as an autosomal recessive genetic trait. Human traits, including the classic genetic diseases, are the product of the interaction of two genes, one received from the father and one from the mother. In recessive disorders, the condition does not appear unless a person inherits the same defective gene for the same trait from each parent. If an individual receives one normal gene and one gene for the disease, the person will be a carrier for the disease, but usually will not show symptoms. The risk of transmitting the disease to the children of a couple, both of whom are carriers for a recessive disorder, is 25 percent. Fifty percent of their children risk being carriers of the disease, but generally will not show symptoms of the disorder. Twenty-five percent of their children may receive both normal genes, one from each parent, and will be genetically normal (for that particular trait). The risk is the same for each pregnancy. Batten Turner Type Congenital Myopathy is an extremely rare disorder that affects males and females in equal numbers. The symptoms of this disorder are most obvious during infancy and childhood. Nine cases of Batten Turner Type Congenital Myopathy have been described in the medical literature. Six of these cases were reported in one family. Adults with Batten Turner Type Congenital Myopathy should be encouraged to get adequate exercise and to avoid obesity. Affected adults may experience episodes of mild muscle weakness, but generally there are no major physical disabilities.